Exam #1 Study Guide

1. All the following are functions of mitochondria, except?

    a. Calcium regulation
    b. Cellular apoptosis
    c. Adenosine triphosphate production
    d. Maintaining cellular membrane potential
    e. All the above are correct

2. True or False – The cristae of the mitochondria are created by infoldings of the outer mitochondrial membrane?

3. The *aconitase* enzyme is an important enzyme in Krebs cycle activity which converts citric acid into isocitric acid. Within the *aconitase* enzyme is a cluster of two elements responsible for proper activation of *aconitase* activity. This same cluster of elements is also found in *succinic acid dehydrogenase*, as well as the electron transport chain. What are the two elements that make up this cluster?

    a. Copper and sulfur
    b. Manganese and iron
    c. Cobalt and copper
    d. Iron and sulfur
    e. None of the above

4. True or False – The amount of ATP produced by the mitochondria in a healthy person is equal to their body weight?

5. How many offspring are fully affected (have the disease) with autosomal recessive inheritance?

    a. 50%
    b. 25%
    c. 75%
    d. 100%
    e. None of the above

6. Which of the following is not correct with regards to mitochondria?

    a. The amount of ATP produced daily from a healthy individual at rest is equal to their body weight.
    b. With maximal activity, the amount of ATP can increase to 1.0 kilogram per minute.
    c. The cells containing mitochondria maintain approximately 250 grams of ATP
    d. Mitochondria are vital to life, but they generally only occupy a small amount of cell volume at approximately 5% to 10%.
    e. Each cell can contain between 1000 to 2500+ mitochondria with the average cell utilizing approximately 10 billion ATP daily.

7. True or False - Most proteins necessary for mitochondrial function are encoded by genes in the cell nucleus and the corresponding proteins are imported into the mitochondrion.

8. *Citrate synthase* is the first enzyme involved in Krebs cycle activity. This enzyme is responsible for producing citric acid by converting which Kreb cycle intermediate, along with its cofactor?

    a. Malic acid and NADH
    b. Succinyl-CoA and GTP
    c. Oxaloacetic acid and Acetyl-CoA
    d. Succinic acid and Fe-S
    e. None of the above

9. True or False – All reactions involving adenosine triphosphate require the presence of magnesium?

10. True or False - In oncology, the Warburg Effect, is the observation that most cancer cells produce energy through a less efficient or "aerobic glycolysis" by glucose uptake, glycolysis, and lactic acid fermentation within the cell despite the presence of oxygen?

11. Anaerobic metabolism accounts for 5% of the cells ATP production, and the Kreb cycle activity accounts for another 5%. How much does the electron transport chain involved in oxidative phosphorylation provide (approximately)?

    a. 10%
    b. 2.5%
    c. 20%
    d. 90%

12. True or False - ATP has no structural similarity to ribonucleic acid (RNA) and deoxyribonucleic acid (DNA) because of the lack of a ribose backbone and phosphate groups.

13. All the following are correct with regards to glycolysis, the production of acetyl-CoA, and enzyme conversion of pyruvate to Acetyl-CoA, except?

    a. Glycolysis is the conversion of glucose to pyruvate. This reaction takes place in the cytosol of the cell where one molecule of glucose (6 carbons) is converted into two 3 carbon pyruvates.

    b. Pyruvate is taken into the mitochondrial where it gets converted to Acetyl-CoA. The enzyme *pyruvate dehydrogenase* (PDH) is responsible for this conversion. The PDH requires various nutrients, e.g., B1, B2, B5, magnesium. It is regulated by another pyruvate enzyme called *pyruvate dehydrogenase kinase* (PDK).

    c. The *pyruvate dehydrogenase* is a complex of different proteins and often referred to as *pyruvate dehydrogenase complex* (PDC). Increase of PDK inhibits PCD activity.

    d. Various cellular compounds limit PDC activity such as Acetyl-CoA, ATP, and NADH.

    e. All the above are correct

14. True or False - The endoplasmic reticulum is considered the energy factory of our cells?

15. What is the innermost aspect of the mitochondria?

    a. Inner mitochondrial membrane

    b. Cristae

    c. Matrix

    d. Outer mitochondrial membrane

    e. None of the above

16. Mitochondrial genes involve sequences for the development of the following electron transport chain (ETC) complexes?

    a. II
    b. I, II, III
    c. I, III, IV, V
    d. V only
    e. None of the above

17. A common problem with certain mitochondrial disorders that affect cellular metabolism is the development of elevated organic acids. Which of the following organic acids, as discussed in module #2, is important and relatively easy to assess through blood testing and/or organic acids testing?

    a. Malonic acid
    b. 2-hydroxyhippuric acid
    c. Lactic acid
    d. 4-hydroxyphenylacetic acid
    e. None of the above

18. This compound which his part of the inner mitochondrial membrane gets released into the intermembrane space triggering apoptosis?

    a. Complex I
    b. ATP Synthetase
    c. Cristae
    d. Cytochrome c
    e. Iron-sulfur cluster

19. True or False – The outer mitochondrial membrane has a protein to phospholipid ratio of 1:1, but the inner mitochondrial membrane is 3:1?

20. True or False - Each ATP molecule is recycled about 1000 times with nearly 70% of the ATP used occurring in the brain and nervous system.

21. All the following are correct regarding cardiolipin as a structural component of the inner mitochondrial membrane (IMM), except?

    a. Cardiolipin makes up about 20% of the total lipid composition of the IMM.
    b. Cardiolipin can be altered in its function by antibodies produced by the immune system.
    c. Cardiolipin contains four phospholipid chains as opposed to two seen with other phospholipids.
    d. Cardiolipin is chemically involved in maintaining optimal cohesiveness of the electron transport chain complexes.
    e. All the above are correct.

22. True or False - mitochondrial disorder refers to primary disorders of mitochondrial metabolism affecting oxidative phosphorylation?

23. All the following are correct with regards to mitochondrial disorders, except?

    a. The prevalence of mitochondrial diseases is estimated to be about 1 in 5000 across all ages.
    b. The clinical manifestations are often complex and can involve all organ systems.
    c. The organ with high energy demands, e.g., brain, eyes, liver, muscles, nerves are particularly at risk.
    d. Phenotypic expression is highly variable in affected members within or between family members, even across the same disease spectrum.
    e. All the above are correct.

24. True or False - The hallmark of mitochondrial disease is the variability of the disease process within the same family or individuals despite similar mutations, which may complicate the diagnosis.

25. Which of the following pairings with regards to mitochondrial disorder symptoms is not correct?

    a. Swallowing - dysphagia
    b. Myopathy - muscle weakness
    c. Ptosis – elevated eye lids
    d. A & B
    e. None of the above

26. True or False – maternal inheritance linked to mitochondrial disorders involves both the man and woman, but mostly the man?

27. Which nucleotides are associated with deoxyribonucleic acid (DNA)?

    a. Adenine
    b. Thymine
    c. Guanine
    d. Cytosine
    e. All the above are correct

28. True or False – The mitochondria have a mechanism for producing heat that does not involve ATP production?

29. Which structure of the mitochondria contains porins?

    a. Matrix
    b. Outer mitochondrial membrane
    c. Cristae
    d. Inner mitochondrial membrane
    e. None of the above

30. True or False - The matrix contains approximately two-thirds of the mitochondrion proteins?