SELF ASSESSMENT - MODULE C: CYSTIC FIBROSIS

1. Which chromosome is involved in the genetic mutation of the gene that causes Cystic Fibrosis? **SEVEN**

2. Cystic Fibrosis is also called **MUCOVISCIDOSIS**.

3. Cystic Fibrosis affects which of the following systems?
   A. Reproductive System
   B. Sweat Glands
   C. Exocrine Glands
   D. Lungs
   E. Pancreas
   F. All the above

4. Patients with cystic fibrosis cannot absorb the fat soluble vitamins which are vitamins (name the vitamins) **A, D, E, K**.

5. Deficiency of vitamin D leads to decreased absorption of **CALCIUM** & **PHOSPHORUS** which is needed for bone growth.

6. A sweat chloride test is positive for CF if the chloride level is greater than **60** mEq/L in children

7. What is meconium ileus? **AN OBSTRUCTION OF THE SMALL INTESTINE OF THE NEWBORN THAT IS CAUSED BY THE IMPACTION OF THICK, DRY TENACIOUS MECONIUM, USUALLY AT OR NEAR THE ILEOCAECAL VALVE.**

8. Cystic Fibrosis patients can be CO_2 retainers True False

9. The most common mutation in CF is known as **ΔF508** which results in the deletion of the amino acid phenylalanine in the CFTR protein.

10. Carriers of a single defective gene (heterozygote) have no clinical disease.
    True False

11. In the Caucasian population, CF is found in 1 out of every **1 IN 3,500** births.

12. CFTR is a protein that regulates **CHLORIDE** movement across epithelial cells.

13. List findings of hyperinflation identified on a chest x-ray.
    A. **Translucent (dark)**
    B. **Depressed or flattened diaphragms**
    C. **Right ventricular enlargement (Cor Pulmonale)**
    D. **Areas of atelectasis and fibrosis**
    E. **Pneumothorax**
    F. **Abscess formation**

14. The term steatorrhea is often used during the physical examination to describe **EXCESSIVE FAT IN THE STOOL**.

15. Name the three following drugs given to patients with cystic fibrosis.

A. Mucolytic used to treat infected sputum **PULMOZYME**.
B. A mucous altering drug used to prevent Na reabsorption into the epithelial cell **AMILORIDE**.
C. An antibiotic **TOBRAMYCIN**.

16. If both parents are carriers of the CF gene, the chance of conceiving a child that has CF is **25%**, is a carrier of the CF gene but does not have CF is **50%**, and is neither a carrier of the CF gene nor has the disease is **25%**.
17. If both parents are carriers of the CF gene, what is the chance of conceiving a child that has CF is **25%**, is a carrier of the CF gene is **50%**, and is neither a carrier of the CF gene nor has the disease is **25%**.

18. Explain the diet necessary for a patient with cystic fibrosis.

- **DIET THERAPY INCLUDES SUFFICIENT CALORIES AND PROTEIN TO PROMOTE NORMAL GROWTH.**
- **A NORMAL-TO-HIGH TOTAL FAT INTAKE TO INCREASE THE CALORIC DENSITY OF THE DIET.**
- **MULTIVITAMINS IN DOUBLE THE RECOMMENDED DAILY ALLOWANCE.**
- **SUPPLEMENTAL VITAMIN E IN WATER-MISCIBLE FORM**
- **SALT SUPPLEMENTATION DURING PERIODS OF THERMAL STRESS AND INCREASED SWEATING.**

19. Why is PEP therapy so beneficial to patients with cystic fibrosis?
PROMOTES SECRETION MOBILIZATION AND PREVENTS AIRWAY COLLAPSE AND RECURRENT PNEUMONIAS