

Alpha-1 Antitrypsin Deficiency and Augmentation Therapy
Prolastin-C Liquid
Post Test

1. Persons with Alpha-1 are at high risk for developing life-threatening liver disease for which of the following reasons?
 - a. The alpha-1 antitrypsin produced by the liver in individuals with Alpha-1 is abnormal.
 - b. The alpha-1 antitrypsin produced by the liver in individuals with Alpha-1 is poorly secreted, resulting in an accumulation within the liver and a marked reduction in circulating alpha-1 antitrypsin protein.
 - c. A and B
 - d. None of the above

2. Persons with Alpha-1 are at high risk for developing life-threatening lung disease for which of the following reasons?
 - a. Low levels of alpha-1 antitrypsin in the lungs allows for the destructive effects of neutrophil elastase to go unchecked resulting in damage to the alveoli and eventually leading to emphysema.
 - b. Accumulation of alpha-1 antitrypsin protein in the lungs damages the gas exchange system of the lungs, the alveoli, and progressively leads to emphysema.
 - c. A and B
 - d. None of the above

3. The greatest single risk factor for Alpha-1 is?
 - a. A positive family history
 - b. Smoking
 - c. Asbestos exposure
 - d. Alcohol abuse

4. Alpha-1 is a genetic disorder in which an abnormal gene is inherited from each parent. Which of the following combinations indicates the four most common genes for Alpha-1?
 - a. M, S, Z and Null
 - b. A, B, D and Null
 - c. Alpha, Beta, Zeta and Null
 - d. None of the above

5. Alpha-1 patients are classified as severely deficient when they have which combination of genes?
 - a. ZZ
 - b. NullNull
 - c. ZNull
 - d. All of the above

6. Lung transplantation is a cure for Alpha-1 Antitrypsin Deficiency.
 - a. True
 - b. False

7. The World Health Organization recommends that all adults with which of the following medical conditions be tested for Alpha-1?
 - a. COPD and/or Chronic Asthma
 - b. Family history of Alpha-1
 - c. Chronic liver disease
 - d. All of the above

8. Which of the following are definitive diagnostic test for Alpha-1?
 - a. Phenotyping, Genotyping, Serum level
 - b. Chest X-ray, CT Scan, Pulmonary Function Tests
 - c. All of the above
 - d. None of the above

9. Which medication noted below, is an FDA approved treatment specifically for Alpha-1 lung disease?
 - a. Serevent
 - b. Albumin
 - c. Progesterone
 - d. Prolastin-C Liquid

10. The criteria for the initiation of Augmentation Therapy include which of the following?
 - a. Patient must be under the care of a licensed physician familiar with the treatment
 - b. Patient should be evaluated for IgA deficiency.
 - c. Epinephrine should be available at initiation of therapy
 - d. All of the above

11. A contraindication for augmentation therapy is?
 - a. Known IgA deficiency
 - b. Penicillin allergy
 - c. Known IgE deficiency
 - d. Egg allergy

12. Which of the following is the recommended dose of Prolastin-C Liquid?
 - a. 80 mg/kg actual body weight once weekly
 - b. 60 mg/kg actual body weight once weekly
 - c. 120 mg/kg actual body weight once weekly
 - d. 60 mg/kg actual body weight bi-weekly

13. Alpha-1 patients are often misdiagnosed for which of the following reasons?
 - a. The usual presenting symptom of Alpha-1 lung disease is dyspnea on exertion
 - b. In existing COPD or asthma patients, the diagnosis of Alpha-1 is not thought of
 - c. Patients with liver cirrhosis are usually suspected of alcohol abuse.
 - d. All of the above

14. A yearly flu vaccination and the pneumonia vaccine every 5-7 years are recommended for patients with Alpha-1.
 - a. True
 - b. False

15. Treatment planning for patients with Alpha-1 lung disease may include which of the following?
 - a. Genetic counseling
 - b. Smoking cessation
 - c. Pulmonary Rehabilitation
 - d. All of the above

16. The products of cigarette combustion are especially destructive to persons with Alpha-1 lung disease for two important reasons, these are:
 - a. Smoking leads to severe halitosis
 - b. Smoke inactivates the alpha-1 antitrypsin in the lungs
 - c. Smoking attracts white blood cells to the lungs in large numbers.
 - d. B and C only

17. Normal levels of alpha-1 antitrypsin may be expressed by two different scales; the micromolar (μM) scale or the milligram per deciliter (mg/dL) scale. Which of the following is a correct value?
 - a. 25-40 mg/dL
 - b. 125-400 μM
 - c. .05-3.0 mg/dL
 - d. 18-40 μM

18. Severe Alpha-1 deficiency is generally identified at what levels?
 - a. Less than 11 μM or less than 58 mg/dL
 - b. Less than 58 μM or less than 11 mg/dL

19. Alpha-1 is the number one genetic cause of which organ transplant in children?
 - a. Lung
 - b. Liver
 - c. Heart
 - d. Kidney

20. Prolastin-C Liquid may be stored
 - a. In the freezer
 - b. At room temperature-not to exceed 77° F for up to 1 month, then must be used or destroyed
 - c. Refrigerated at 2-8° C; 36-46° F
 - d. B and C only

21. Universal Precautions and strict aseptic technique are to be instituted:
 - a. Only during IV catheter placement
 - b. Only during the removal of the IV catheter
 - c. During pooling of the drug
 - d. During all drug preparation and administration activities

22. Patients receiving augmentation therapy with Prolastin-C Liquid can expect that with every infusion, the same number of vials will be the same to achieve their prescribed dose.
 - a. True
 - b. False

23. Prolastin C Liquid should be administered using an inline 15-micron filter.
- True
 - False
24. Vials of Prolastin C Liquid are produced in what size:
- 5000 mg vial containing 50 ml of solution for injection
 - 1000 mg vial containing 20 ml of solution for injection
 - 500 mg vial with 20 ml of solution for injection
 - All the above
 - None of the above
25. In general, a weekly infusion takes approximately 2-3 hours to infuse.
- True
 - False
26. Indicate the correct infusion rate of Prolastin-C Liquid for a patient weighing 210 lbs., receiving a weekly dose of 60mg/kg and the IV tubing drop factor = 10gtts/ml.
- 5700 mgs
 - 1260 mgs
 - 335 ml/hr
 - 456 ml/hr
27. Known side effects of Prolastin-C Liquid include, but are not limited to:
- Sun sensitivity, psoriasis and renal failure
 - Nausea, vomiting and bloody diarrhea
 - Chills, malaise, headache and rash
 - Cough, hoarseness and increased sputum production
28. Augmentation therapy is self-limiting. Once the patient's symptoms improve, therapy is discontinued.
- True
 - False
29. Prolastin-C Liquid was the first FDA approved therapy specifically for the treatment of severe Alpha-1 Antitrypsin Deficiency and has been available since 1987.
- True
 - False
30. The goal of augmentation therapy with Prolastin-C Liquid is to:
- Restore the circulating level of alpha-1 protein toward normal
 - Normalize LFT's
 - Restore lung function
 - Raise serum levels of trypsin
31. All equipment used in the process of pooling Prolastin-C Liquid should be discarded in the household trash.
- True
 - False

32. Prolastin-C Liquid should appear prior to administration:
- Red tinged in solution
 - Clear to straw colored
 - Dark amber
 - Cloudy
33. It is expected that particulates will be seen in Prolastin-C Liquid.
- True
 - False
34. Alpha-1 Antitrypsin Deficiency was first recognized in the early 1960's by?
- Professors Laurel and Hardy
 - Dr. Smith and Will Robinson
 - Professor Carl-Bertil Laurell and Dr. Sten Eriksson
 - Dr. David Livinstone
35. Alpha-1 Antitrypsin Deficiency is more than twice as common of which other genetic disorder?
- Gaucher's Disease
 - Tay-Sack's Disease
 - Cystic Fibrosis
 - Huntington Disease
36. It is believed that Alpha-1 affects as many as 100,000 individuals in the United States.
- True
 - False
37. Panniculitis is a painful inflammatory skin disorder sometimes associated with Alpha-1 Antitrypsin Deficiency?
- True
 - False
38. Aspects of the treatment plan for individuals with Alpha-1 includes:
- Smoking cessation and avoidance of environmental pollutants
 - Prompt treatment of respiratory infections, proper nutrition and exercise
 - A and B
 - Annual gastroscopy
39. Resources in the community to assist those affected by Alpha-1 Antitrypsin Deficiency includes which of the following?
- The Alpha-1 Foundation
 - The American Lung Association
 - AlphaNet
 - All of the above
40. It is important to establish IV access prior to pooling Prolastin-C Liquid because:
- It is good nursing practice when administering IV medications in the home
 - Prolastin-C Liquid is expensive and if IV access can not be established, the product may be inadvertently wasted
 - Prolastin-C Liquid contains no preservative and must be infused within 3 hrs
 - All of the above