

CHAPTER 16. CYSTIC FIBROSIS, SELF-ASSESSMENT QUESTIONS

1. Airway clearance therapy in patients with cystic fibrosis (CF) should be performed by the patient or patient's caregiver:

- A. Only when the patient is symptomatic
- B. At least three times per day during acute exacerbations
- C. Every Monday, Wednesday, and Friday
- D. Only under the direct supervision of a respiratory therapist
- E. With at least two different methods

2. A 5-year-old male CF patient presents with poor nutritional status. Height is 105 cm (25th percentile), and weight is 14.5 kg (< 3rd percentile). He takes Zenpep 10,000 two caps with meals and one cap with snacks, and he has four loose stools per day. Which of the following are appropriate nutritional interventions?

- A. Dietary counseling to increase caloric intake
- B. Increase Zenpep 10,000 to three caps with meals and two caps with snacks
- C. Admit to hospital for initiation of intravenous parenteral nutrition
- D. A and B
- E. A, B, and C

3. Dornase alfa is used in CF for which of the following reasons:

- A. Chronic *Pseudomonas aeruginosa* suppression
- B. Mucolytic activity
- C. Sputum induction
- D. A and B
- E. B and C

4. The following culture and sensitivity results are reported for a 13-year-old CF patient:

Current month: *P. aeruginosa* (sensitive to cefepime, ceftazidime, piperacillin-tazobactam, meropenem, aztreonam, gentamicin, tobramycin, amikacin, ciprofloxacin).

One month ago: *S. aureus* (sensitive to vancomycin, sulfamethoxazole-trimethoprim, minocycline, linezolid; resistant to cefazolin, clindamycin, erythromycin, nafcillin, gentamicin) and *P. aeruginosa* (sensitive to cefepime, ceftazidime, piperacillin-tazobactam, meropenem, aztreonam, tobramycin, amikacin; resistant to gentamicin, ciprofloxacin)

Six months ago: *A. xylosoxidans* (sensitive to ceftazidime, sulfamethoxazole-trimethoprim, and piperacillin-tazobactam; resistant to ticarcillin-clavulanate, minocycline, levofloxacin)

Based on these results, the most appropriate inpatient antibiotic regimen is:

- A. Piperacillin-tazobactam, ciprofloxacin, and vancomycin
- B. Piperacillin-tazobactam, tobramycin, and vancomycin
- C. Cefepime, tobramycin, and clindamycin
- D. Cefepime, tobramycin, and minocycline
- E. Ceftazidime, gentamicin, and sulfamethoxazole-trimethoprim

5. Patients with a history of anaphylactic reaction to ticarcillin-clavulanate and cefepime can most safely be treated with which antibiotic?

- A. Meropenem
- B. Ertapenem
- C. Loracarbef
- D. Aztreonam
- E. Dicloxacillin

6. An 8-year-old CF patient with *F508del* and *G551D* CFTR gene mutations is prescribed ivacaftor for chronic maintenance therapy. Which of the following statements should *not* be included in medication counseling?

- A. Ivacaftor interacts with many medications. Ask your CF doctor or pharmacist before stating any new medications.
- B. All other chronic maintenance therapies should be continued after starting ivacaftor.
- C. Ivacaftor should be taken on an empty stomach.
- D. Blood tests will be ordered periodically to monitor liver function while taking ivacaftor.
- E. Adherence to therapy is very important to optimize the effectiveness of ivacaftor.

7. CF patients have altered pharmacokinetic parameters because of:

- A. Increased fat stores
- B. Chronic maldigestion leading to increased protein binding
- C. Hepatic sequestration of antibiotics
- D. Increased percent lean body mass compared with total body mass
- E. Tolerance of faster infusion rates

8. The preferred treatment for a patient with newly diagnosed cystic fibrosis–related diabetes (CFRD) without documented fasting hyperglycemia is:

- A. Insulin glargine 20 units subcutaneously every evening
- B. Lispro insulin 1 unit per 15 g of carbohydrates with meals and snacks
- C. Isophane insulin sliding scale for glucose levels greater than 150 mg/dL (8.3 mmol/L)
- D. Metformin 500 mg orally twice daily
- E. Glyburide 5 mg orally once daily

9. Current maintenance therapy for CF is designed with the following goal(s) in mind:

- A. Delay of disease progression
- B. Normal growth and development
- C. Disease cure
- D. Two of the above
- E. All of the above

10. A 25-year-old CF patient (weight: 48 kg) with a history of *P. aeruginosa* colonization presents to clinic with complaints of slowly declining lung function over the past 2 years. FEV₁ has declined from 62% predicted to 48% predicted during this time. Current maintenance therapies include albuterol 2.5 mg nebulized twice daily with chest physiotherapy, dornase alfa 2.5 mg nebulized every morning, hypertonic saline 7% 4 mL nebulized every evening, AquADEKs one capsule daily, omeprazole 20 mg daily, and CREON 24,000 three caps with meals and two caps with snacks. What antiinflammatory treatment is most appropriate for this patient?

- A. Azithromycin 500 mg three times per week
- B. Ibuprofen 20 mg/kg/dose twice daily
- C. Fluticasone 220 mcg two puffs twice daily
- D. Prednisone 10 mg daily
- E. All of the above are reasonable treatment options

11. A 48-year-old female patient (weight: 38 kg) with a history of CF diagnosed at age 5 is admitted through the emergency department for treatment of a pulmonary exacerbation. Her FEV₁ is 21% predicted. Additional medical history is significant for CFRD, osteoporosis, pancreatic insufficiency, depression, and pneumothorax. The patient is new to your CF center, and no pharmacokinetic history is available. Which of the following statements is true?

- A. The patient is likely to have increased tobramycin clearance due to pancreatic insufficiency
- B. Serum creatinine does not need to be screened in this patient
- C. Tobramycin should be dosed every 8 hours in all CF patients
- D. This patient is at higher risk for delayed tobramycin clearance due to age and cumulative lifetime exposure to aminoglycosides
- E. Aminoglycosides should be avoided in this patient because pharmacokinetic history is unknown

12. Which of the following scenarios is most likely to cause pancreatic enzyme replacement therapy treatment failure?

- A. Opening enzyme capsules and sprinkling beads on applesauce
- B. Crushing enzyme beads and mixing in with infant formula
- C. Taking enzyme capsules throughout a meal
- D. Eating an apple every afternoon without taking enzymes
- E. Taking ranitidine as needed for heartburn symptoms

13. A 21-year-old CF patient wishes to start chronic suppressive antibiotic therapy due to frequent exacerbations and declining lung function. Culture history is notable for MRSA, *P. aeruginosa*, and *Aspergillus fumigatus*. Which of the following is not a favorable option?

- A. Aztreonam lysine inhalation 75 mg three times daily, alternating cycles of 28 days on/off
- B. Tobramycin inhalation powder 112 mg twice daily, alternating cycles of 28 days on/off
- C. Colistin 150 mg inhalation twice daily, alternating cycles of 28 days on/off
- D. Minocycline 100 mg by mouth twice daily with alternating inhaled tobramycin solution 300 mg BID in 28-day cycles
- E. All of the above are routinely accepted regimens and should be selected by patient preference

14. Patients on chronic azithromycin therapy should be monitored for:

- A. Growth of *Mycobacterium abscessus* on sputum cultures
- B. Development of renal toxicity
- C. Hyperglycemia
- D. Osteoporosis
- E. Thrombocytopenia

15. CF patients on IV aminoglycosides for pulmonary exacerbations should be monitored by which of the following methods?

- A. Daily tobramycin trough concentrations
- B. Peak and trough concentrations weekly
- C. Initial determination of peak and trough concentrations per local CF center protocol, then weekly trough concentrations
- D. Initial determination of peak and trough concentrations per local CF center protocol, then weekly peak concentrations
- E. No concentration monitoring is needed with empiric dosing as long as serum creatinine is within normal limits for age

ANSWERS

- 1. B
- 2. D
- 3. B
- 4. B
- 5. D
- 6. C

7. D

8. B

9. D

10. A

11. D

12. B

13. D

14. A

15. C