

Chapter 68, Self-Assessment Questions

1. Which of the following factors can contribute to increased risk for vasoocclusion in sickle cell disease patients?
 - A. Decreased red blood cell (RBC) deformability
 - B. High RBC fetal hemoglobin (HbF) concentration
 - C. Low RBC adult hemoglobin (HbA) concentration
 - D. Rapid RBC transit through microcirculation
 - E. Decreased RBC viscosity

2. MM is a 9-year-old boy with sickle cell disease (SCD) who has just had a stroke. He is starting to regain some motor function and is progressing well. What is the most effective measure that can be taken to prevent further strokes in MM?
 - A. Hydroxyurea taken daily for life
 - B. IV antibiotics for 10 days to prevent meningitis
 - C. Anticoagulant medications
 - D. Chronic transfusion RBC therapy for life
 - E. Iron chelation therapy

3. Which of the following choices may *best* help to prevent infectious complications from sickle cell disease?
 - A. Immunize for *S. pneumoniae* and take daily oral PenVK until age 6
 - B. Immunize for *H. flu* and take oral PenVK at the first sign of a fever
 - C. Immunize for Hepatitis B and give IV antibiotics at the first sign of a fever
 - D. Immunize for influenza and keep the patient adequately hydrated

- E. Immunize for *N.meningitidis* and take daily oral amoxicillin until age 6
4. A 30-year old man with sickle cell disease presents to the ER with complaints of lethargy and tiredness increasing over the past 2 weeks. His oxygen saturation level is 86% (0.86) on room air. Hemoglobin is 5.4 g/dL (54 g/L; 3.35 mmol/L). WBC is $5 \times 10^3/\text{mm}^3$ ($5 \times 10^9/\text{L}$), platelets $100 \times 10^3/\text{mm}^3$ ($100 \times 10^9/\text{L}$). Oral temperature is 37.5°C. What *one* therapeutic intervention will have the *best* impact on his chief complaint?
- A. IV antibiotics
 - B. Packed RBC transfusion
 - C. IV fluids (give two times maintenance)
 - D. Oxygen to keep saturations > 95% (0.95)
 - E. IV morphine
5. A 22-year-old woman with SCD is admitted for acute chest syndrome. She has an oral temperature of 102.2°F (39.0°C), is receiving appropriate fluid, and is receiving oxygen by nasal cannula because her oxygen saturations on room air were 85% (0.85). Her drug profile includes intravenous morphine for pain and oral diphenhydramine for itching. Which one of the following is most appropriate to add to this patient's regimen at this time?
- A. Methylprednisolone IV every 12 hours.
 - B. Albuterol metered-dose inhaler every 4 hours.
 - C. Ceftriaxone IV every 12 hours.
 - D. Vancomycin IV every 12 hours
 - E. Hydroxyurea orally every 24 hours
6. A 2-year-old child with SCD should receive which of the following in addition to the required childhood immunizations?

- A. Influenza vaccine
 - B. Meningococcal vaccine
 - C. Papillomavirus vaccine
 - D. PPV 23 vaccine
 - E. Rotavirus vaccine
7. A child who presents with severe anemia due to sequestration crisis should receive IV fluids and:
- A. Decitabine
 - B. Epoetin
 - C. Hydroxyurea
 - D. Morphine
 - E. RBC transfusion
8. Select the appropriate penicillin prophylaxis regimen for a 4-year-old child with SCD
- A. 250 mg twice daily until 5 years of age, then discontinue
 - B. 250 mg twice daily and continue through adolescence
 - C. 125 mg twice daily until 5 years of age, then discontinue
 - D. 125 mg twice daily and continue through adolescence
 - E. 125 mg twice daily until 5 years of age, then 250 mg twice daily
9. Which laboratory parameter necessitates temporary discontinuation of hydroxyurea therapy in a 14-year-old boy?
- A. Absolute neutrophil count less than $7 \times 10^3/\mu\text{L}$ ($7 \times 10^9/\text{L}$)
 - B. Hemoglobin value less than 5 g/dL (50 g/L or 3.1 mmol/L)
 - C. Mean corpuscular volume less than 72 fL ($72/\mu\text{m}^3$)

- D. Platelets less than $90 \times 10^3/\mu\text{L}$ ($90 \times 10^9/\text{L}$)
- E. Reticulocytes less than $80 \times 10^3/\mu\text{L}$ ($80 \times 10^9/\text{L}$)
10. All of the following are complications that can occur because of vasoocclusion *except*:
- A. Congestive heart failure
 - B. Leg ulcers
 - C. Priapism
 - D. Renal insufficiency
 - E. Stroke
11. Which of the following therapies is an appropriate treatment for SCD-related priapism?
- A. Corticosteroids, such as prednisone
 - B. Antibiotics, such as ceftriaxone
 - C. Vasoconstrictors, such as epinephrine
 - D. IV fluids, such as D51/2NS
 - E. All of the above
12. RS is a 20-year old man with SCD who takes folic acid daily along with ibuprofen and hydrocodone/acetaminophen at least 4 days a week to control his baseline pain. He presents to the ED with right shoulder pain that he rates as 9/10 on a visual analog pain scale. He is to be admitted to the hospital for pain control. What is the most appropriate initial pain medication choice for this patient?
- A. Continue his ibuprofen and hydrocodone/acetaminophen on a scheduled basis
 - B. Start ketorolac IV every 6 hours with scheduled oral morphine
 - C. Start acetaminophen IV every 6 hours with scheduled IV meperidine
 - D. Start ketorolac IV every 6 hours with morphine PCA

- E. Start ibuprofen IV every 6 hours with fentanyl PCA
13. Select the most appropriate empiric treatment for suspected pneumonia in an adult patient with SCD.
- A. Cefotaxime + vancomycin
 - B. Ciprofloxacin + erythromycin
 - C. Gentamicin + ampicillin
 - D. Meropenem + penicillin
 - E. Piperacillin/tazobactam + tobramycin
14. TL is a 9-year-old girl with SCD who has been admitted to the hospital for pain control associated with a vasoocclusive crisis. She is currently receiving a morphine PCA with both a continuous basal rate as well as on-demand dosing. Today on hospitalization day 3, she is complaining of severe itching all over, which is distressing to her. Which of the following is the best option for treating her pruritus?
- A. Triamcinolone 0.1% applied topically to the affected area every 6 hours.
 - B. Diphenhydramine cream 1% applied topically to the affected area every 6 hours.
 - C. Hydroxyzine intramuscularly every 6 hours as needed.
 - D. Diphenhydramine intravenously every 6 hours.
 - E. Loratadine orally every 24 hours
15. A 3-year-old girl (Wt 13.5 kg) with SCD presents with a serum ferritin concentration of 2500 ng/mL (2500 mcg/L; 5618 pmol/L) because of frequent RBC transfusions. Which of the following statements concerning deferasirox (Exjade) is the best when deciding the *initial* intervention for this patient?
- A. Deferasirox is not indicated in young children

- B. Give 250-mg tablet once daily
- c. Give 250-mg tablet + 125-mg tablet once daily
- D. Give 500-mg tablet once daily
- E. Give 500-mg tablet + 125-mg tablet once daily

Answers

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