**Clinical Case Segment- April StuNews**

**Reviewed by ACCP Clinical Pharmacy Challenge Exam Panel**

Vignette: A 4-year-old boy with sickle cell disease presents to urgent care with mild chest pain. His mother states that he has had a productive cough for the past week with increasing frequency and low-grade fever. His pain score today is 4/10 using the Wong-Baker FACES pain scale.

Past Medical History: Sickle cell disease (HbSS), hospitalizations for pain crises (once or twice yearly), previous hospitalizations for pneumonia (including one pediatric intensive care unit admission) in the past 12 months, moderate persistent asthma, allergic rhinitis, gastroesophageal reflux disease, splenectomy (age 3 years)

Social History: Lives with his mother and father (adopted), presently in first grade

Current Medications: Fluticasone 110 mcg HFA (hydrofluoroalkane) metered-dose inhaler 2 puffs inhaled twice daily with spacer; cetirizine 10 mg daily; fluticasone nasal spray 1 spray each nostril daily; montelukast 5 mg daily; hydroxyurea 500 mg daily; lansoprazole 15 mg daily; hydrocodone/acetaminophen 5 mg/325 mg every 4–6 hours as needed; ibuprofen 200 mg every 6 hours as needed for fever

Allergies: No known drug allergies

Vital Signs: BP 92/45 mm Hg; HR 90 beats/minute; Temp 100.6°F (38.1°C); RR 22 breaths/minute; height 115 cm; weight 25 kg

Laboratory Values: Spo2 91% on room air; basic metabolic panel within normal limits

WBC 16.5 x 103 cells/mm3 (16.5 x 109/L); RBC 3.1 x 106 cells/mm3 (3.1 x 1012/L); Hgb 8.8 g/dL (88 g/L); Hct 30% (0.3); MCV 100 fL/cell; MCH 35 pg/cell; MCHC 35 g/dL; red cell distribution width 18%; Plt 200,000/mm3 (200 x 109/L); mean platelet volume 9.2 fL; neutrophils 70%; lymphocytes 22%; monocytes 3%; eosinophils 5%; reticulocyte count 10; anisocytosis 1+; sickle cells 2+

Procedures: Chest radiograph reveals lower left lobe opacity with consolidation.

Other: Immunization History: Parents have declined vaccines

**Question 1**

Which pathogen is most likely the cause of the patient’s pneumonia?

1. *Achromobacter xylosoxidans*
2. *Escherichia coli*
3. *Pseudomonas aeruginosa*
4. *Streptococcus pneumoniae*

Answer: 4. *S. pneumoniae*

Rationale: Because the patient is asplenic, has an unknown immunization status, and has a sickle cell condition, he is at increased risk of infection because of encapsulated bacteria such as *S. pneumoniae*. The other pathogens are less likely.

Citations:

Hagemann TM. Pediatric sickle-cell disease. In: Benavides S, Nahata MC, eds. Pediatric Pharmacotherapy. Lenexa, KS: American College of Clinical Pharmacy, 2013:785-95.

Yawn BP, Buchanan GR, Afenyi-Annan AN, et al. Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. JAMA 2014;312:1033-48.

**Question 2**

Which medication is likely to cause neutropenia in this patient?

1. Hydroxyurea
2. Ibuprofen
3. Lansoprazole
4. Montelukast

Answer: 1. Hydroxyurea

Rationale: Myelosuppression (e.g., neutropenia, thrombocytopenia, anemia) is common with hydroxyurea. Because ibuprofen is used as needed for fever, it is less likely to be the cause of the patient’s neutropenia. Neutropenia with the use of lansoprazole is rare and not reported with montelukast.

Citation: Hagemann TM. Pediatric sickle-cell disease. In: Benavides S, Nahata MC, eds. Pediatric Pharmacotherapy. Lenexa, KS: American College of Clinical Pharmacy, 2013:785-95.

**Question 3**

Which supplement would most likely address chronic hemolysis caused by sickle cell disease?

1. Ascorbic acid
2. Cholecalciferol
3. Folic acid
4. Magnesium oxide

Answer: 3. Folic acid

Rationale: In children with chronic hemolysis caused by sickle cell disease, the addition of folic acid supplementation may be beneficial. Folic acid replenishes the depleted folate stores necessary for erythropoiesis. Folic acid supplementation is well established in the treatment of chronic hemolytic anemia. Current guidelines recommend supplementation of 1 mg daily. The other supplements have not been shown to reduce hemolysis.

Citations:

Hagemann TM. Pediatric sickle-cell disease. In: Benavides S, Nahata MC, eds. Pediatric Pharmacotherapy. Lenexa, KS: American College of Clinical Pharmacy, 2013:785-95.

van der Dijs FP, Fokkema MR, Dijck-Brouwer DA, et al. Optimization of folic acid, vitamin B(12), and vitamin B(6) supplements in pediatric patients with sickle cell disease. Am J Hematol 2002;69:239-46.

**Question 4**

The medical resident asks for a recommendation regarding antibiotic prophylaxis in this patient. Which is the MOST appropriate response?

1. Because the patient is older than 3 years, prophylaxis is not required.
2. If he receives the pneumococcal vaccination, prophylaxis is not required.
3. Prophylaxis is indicated. Initiate amoxicillin 250 mg twice daily.
4. Prophylaxis is indicated. Initiate penicillin 250 mg twice daily.

Answer: 4. Prophylaxis is indicated. Initiate penicillin 250 mg twice daily.

Rationale: Prophylactic penicillin is recommended in all children with sickle cell disease until age 5 years (Strong Recommendation). Because this patient is younger than 5, prophylaxis with penicillin 250 mg twice daily is recommended by the guidelines (option 4 is correct). A dose of 125 mg twice daily is used for children younger than 3 years (option 1 is incorrect). Regardless of his pneumococcal vaccination status, this patient should continue to receive penicillin prophylaxis until at least age 5 (option 2 is incorrect). Amoxicillin is not indicated for prophylaxis in this patient population (option 3 is incorrect).

Citations:

Hagemann TM. Pediatric sickle-cell disease. In: Benavides S, Nahata MC, eds. Pediatric Pharmacotherapy. Lenexa, KS: American College of Clinical Pharmacy, 2013:785-95.

Yawn BP, Buchanan GR, Afenyi-Annan AN, et al. Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. JAMA 2014;312:1033-48.

**Question 5**

The patient later requires repeated transfusions because of his sickle cell disease. After 18 months of transfusions, he needs treatment for iron overload. Which agent will help iron chelation with deferoxamine treatment?

1. Ascorbic acid
2. Calcium carbonate
3. Lansoprazole
4. Vitamin B12

Answer: 1. Ascorbic acid

Rationale: Patients who are treated with deferoxamine should have ascorbic acid added to their regimen because it will help increase the availability of iron for chelation. The other agents listed do not affect the availability of iron for chelation by deferoxamine.

Citation: Hagemann TM. Pediatric sickle-cell disease. In: Benavides S, Nahata MC, eds. Pediatric Pharmacotherapy. Lenexa, KS: American College of Clinical Pharmacy, 2013:785-95.