

A 3-year-old boy with sickle cell anemia is brought to the emergency department due to fever. He has chills but no bone pain, cough, or difficulty breathing. The patient's medical history includes hospitalization as an infant for dactylitis and again last summer for splenic sequestration crisis. Other than ibuprofen and acetaminophen for fever and pain, he takes no medications regularly and his vaccinations are current. Temperature is 39.5 C (103.2 F), blood pressure is 78/40 mm Hg, pulse is 140/min, and respiratory rate is 22/min. Physical examination shows a lethargic boy with mild pallor. Laboratory results are as follows:

Hematocrit	24%
Platelets	325,000/mm ³
Leukocytes	18,800/mm ³
Neutrophils	80%
Bands	13%

Blood culture is pending. Which of the following organisms is the most likely cause of this patient's condition?

- ☐ A. *Haemophilus influenzae* type B
- ☐ B. *Neisseria meningitidis*
- ☐ C. *Pseudomonas aeruginosa*
- ☐ D. *Salmonella enteritidis*
- ☐ E. *Staphylococcus aureus*
- ☐ F. *Streptococcus pneumoniae*

Submit

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- ☐ A. *Haemophilus influenzae* type B [5%]
- ☐ B. *Neisseria meningitidis* [6%]
- ☐ C. *Pseudomonas aeruginosa* [3%]
- ☐ D. *Salmonella enteritidis* [15%]
- ☐ E. *Staphylococcus aureus* [14%]
- ☒ F. *Streptococcus pneumoniae* [56%]

Proceed to Next Item

Explanation:

User Id: [REDACTED]

Sickle cell anemia	
Pathophysiology	<ul style="list-style-type: none">• B-globin mutation• Autosomal recessive inheritance

Explanation:

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Sickle cell anemia	
Pathophysiology	<ul style="list-style-type: none">• B-globin mutation• Autosomal recessive inheritance
Clinical features	<ul style="list-style-type: none">• Hemolytic anemia• Vaso-occlusive pain crises
Laboratory findings	<ul style="list-style-type: none">• ↓ Hematocrit, ↑ reticulocytes• Peripheral smear: Sickled red blood cells, Howell-Jolly bodies
Management	Maintenance <ul style="list-style-type: none">• Vaccination• Penicillin (until age 5)• Folic acid supplementation Acute pain crises <ul style="list-style-type: none">• Hydration• Analgesia• ± Transfusion

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This patient's fever, chills, hypotension, leukocytosis, and bandemia are consistent with sepsis. Intermittent sickling of red blood cells in the spleen leads to splenic infarction and ultimately **functional asplenia**. As a result, patients with sickle cell disease (SCD) are at high risk for sepsis from *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Neisseria meningitidis*.

The incidence of bacteremia has decreased as a result of improved **vaccination** approaches (eg, pneumococcal conjugate and polysaccharide vaccines, meningococcal conjugate vaccine). Despite vaccination, *S pneumoniae* remains by far the most common cause of sepsis in patients with SCD, usually from **non-vaccine serotypes**. Therefore, patients with SCD should receive prophylactic **penicillin** until at least age 5. Unfortunately, this patient was not taking penicillin.

- \pm Transfusion

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(Choices A and B) *H influenzae* and *N meningitidis* are encapsulated organisms that can cause sepsis in patients with SCD; however, they are extremely rare in vaccinated patients. In contrast, *S pneumoniae* remains a common cause of sepsis despite immunization due to non-vaccine serotypes.

(Choice C) *Pseudomonas aeruginosa* is a common cause of bacteremia in patients with burn wounds and neutropenia. It is an unusual cause of sepsis among patients with SCD as the majority of *P aeruginosa* isolates are not encapsulated.

(Choices D and E) *Salmonella* species and *Staphylococcus aureus* are the 2 most common causes of osteomyelitis in patients with SCD. However, this patient has no bone pain suggestive of osteomyelitis, and neither organism is a common cause of sepsis.

Educational objective:

Sickle cell disease (SCD) causes functional asplenia due to recurrent splenic infarction. Therefore, patients are at risk of overwhelming infection with encapsulated organisms and should receive vaccination and penicillin prophylaxis. Pneumococcus remains the most common cause of sepsis in patients with SCD.

References:

1. **Pneumococcal bacteremia in a vaccinated pediatric sickle cell disease population.**
2. **Bacteremia in children with sickle hemoglobinopathies.**
3. **Increase in invasive *Streptococcus pneumoniae* infections in children with sickle cell disease since pneumococcal conjugate vaccine**

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