1.0 Introduction

By 3–4 months of age (when fetal hemoglobin declines to <50% of total), many children with sickle cell anemia (HbSS) and sickle β-thalassemia develop clinically significant hemolytic anemia and impairment of splenic function. In others, although the HbF may remain above 50% these children are still at risk of splenic hypofunction. Even though the spleen may be enlarged during the first years of life, its phagocytic function is markedly reduced. Therefore, children with sickle cell anemia are at risk of overwhelming septicemia, often without a primary focus, due to encapsulated organisms, including Streptococcus pneumonia and Haemophilus influenza type B.

This clinical practice guideline has been developed for the management of febrile patients with sickle cell disease who present to the emergency department or are inpatients.

2.0 Preventative Management

• To reduce high mortality, we strongly recommend:
  
  • Early diagnosis of sickle cell anemia by newborn screening and referral to a comprehensive care program for sickle cell disease. With newborn screening in place since November 2006, patients should be seen within 3 months of birth.
  
  • Prophylactic penicillin or amoxicillin, to be prescribed as soon as sickle cell disease is diagnosed, and continued until at least 5 years of age (to be continued past the age of 5 years in certain circumstances). In patients with significant beta-lactam allergy, trimethoprim-sulfamethoxazole should be used.
  
  • Vaccination against pneumococcus, meningococcus and haemophilus influenza type B. Annual influenza vaccine is also recommended.

• Despite these measures, septicemia may still occur. Whenever a child with sickle cell disease has an oral or rectal temperature >38.5°C or an axillary temperature >38°C, he or she should be seen urgently. Febrile young infants (<3 months of age) should have an appropriate infectious work up, irrespective of their sickle cell status.
3.0 Clinical Recommendations for Management of Fever in Patients with Sickle Cell Disease

Emergency Department Initial Assessment and Management:
1. Complete history, physical exam, and additional investigations using ED Sickle Cell/Fever order set
2. Consider admission to inpatient unit if criteria below are met:
   - Temperature > 38.5°C or axillary temperature > 38°C
   - Symptoms suggesting meningitis
   - Presence of jaundice or pallor
   - Clinical suspicion of acute chest syndrome
3. Obtain CBC, differential, reticulocyte count, sodium, potassium, creatinine, liver enzymes, and chest x-ray if indicated
4. Consider admission to inpatient unit if lab results are abnormal
5. If patient is neutropenic, consult Hematology on call prior to discharge
6. Refer to Infectious Disease for patient with meningitis
7. Refer to Preventive Medicine for patient with osteomyelitis
8. Refer to Neonatal Intensive Care Unit for patient with sepsis
9. Discharge patient home per ED Sickle Cell/Fever order set

Emergency Department Monitoring
- Monitor vital signs
- Observe closely for deterioration in clinical status
- Administer Tylenol and Ibuprofen
- Provide dose of oral antibiotics for patient with meningitis
- Consider antibiotics if patient presents with cough, chest pain, hypoxia, or clinical suspicion for pneumonia/acute chest syndrome
- Consult Hematology for communication on the development of chest crisis
- Observe for development of chest crisis
- Consider admission to inpatient unit if lab results are abnormal

Initiate inpatient Sickle Cell order set in Epic
- Document patient’s admission to Sickle Cell service
- Document patient’s fever and symptoms
- Document patient’s laboratory results
- Document patient’s management

Consent to discharges
- Document patient’s discharge from hospital
- Discharge patient home with appropriate follow-up
- Provide discharge instructions
- Provide patient education
- Provide patient’s family education
- Provide patient’s family education
- Provide patient’s follow-up care

Printable Version

4.0 References


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**5.0 Related documents**

- [Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease](#)
- [Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease](#)

**Attachments:**

- [Fever Care Pathway Final 2021.pdf](#)
- [Revision History.docx](#)
- [SC_Clinic Follow Up Revised 2021_FINAL.pdf](#)
- [SCD fever_criteria for admission 2021 FINAL.pdf](#)
- [SCD fever_discharge planning process 2021 FINAL.pdf](#)
- [SCD fever_inpatient management.pdf](#)
- [SCD fever_out patient follow up.pdf](#)
- [SCD pain plan_july 2015.pdf](#)