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:
- Complete history, physical exam, and multispecialty investigations using ED Sickle Cell Fever order set.
- Consider hospitalization for children with SCD, especially for high-risk groups (e.g., splenic sequestration, basilar pneumonia), to manage complications such as sepsis and meningitis in children with sickle cell disease.
- Notify hematology fellow on call prior to discharge home from ED.
- Follow up with primary care provider for follow up and discharge planning process.

Emergency Department Monitoring:
- Monitor vital signs every hour (e.g., respiratory rate, oxygen saturation, blood pressure, heart rate).
- Monitor for complications such as sepsis and meningitis, especially in children with sickle cell disease.
- Refer to Acute Painful Episodes Vaso Ray (APECED) for additional investigations.

Inpatient Care:
- Tolerating fluids and medications by mouth.
- Initiating inpatient Sickle Cell order set in Epic.
- Outpatient management of febrile illness in infants and young children with sickle cell disease.

Inpatient Management:
- Management of fever in children with SCD and complications such as meningitis.
- Administration of antipyretics and antibiotics as per institutional protocol.
- Follow up with primary care provider for follow up and discharge planning process.

4.0 References


### 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](#)