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 hypo function. 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 routine blood work using ED Sickle Cell Fever order set.
- Consider admission of all children with oral or rectal temperature > 38.5ºC as ACS is common in SCD patients presenting to ED.
- Obtain blood culture on all children with fever.
- Consider obtaining chest X-ray if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.
- Call Hematology for all patients with the following: fever, bacterial infection, or otherwise concerned about the patient's clinical status.

**Emergency Department Monitoring:**

- Ensure patient meets the criteria for SCD (SCD) presents to ED with fever (oral or rectal temperature > 38.5ºC or axillary > 38ºC) – obtain if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.
- Provide dose of oral antibiotics for UTI or known renal anomaly – obtain if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.
- Initiate inpatient Sickle Cell order set in Epic.

**Follow-up confirmed by telephone before discharge:**

- Tylenol and Ibuprofen – initiate inpatient management document.
- Follow-up confirmed by telephone before discharge.
- Complete history and physical exam including growth chart, complications of SCD, previous admissions, medications, surgical history.
- FBC, differential, reticulocyte count, sodium, potassium, creatinine, liver function tests, ESR, ACST.
- Obtain chest X-ray if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.
- Consult Hematology service for all patients with the following: fever, bacterial infection, or otherwise concerned about the patient's clinical status.
- Patient's discharge and transition plan – discharge planning process.

**4.0 References**


©The Hospital for Sick Children (“SickKids”), All Rights Reserved. This document was developed solely for use at SickKids. SickKids accepts no responsibility for use of this material by any person or organization not associated with SickKids. A printed copy of this document may not reflect the current, electronic version on the SickKids Intranet. Use of this document in any setting must be subject to the professional judgment of the user. No part of the document should be used for publication without prior written consent of SickKids.


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](#)