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 additional investigations using SickKids Sickle Cell Fever order set.
- Neonates (0-28 days of age) need a minimum of 2 visits to the emergency department. 2-3 weeks of age visit is recommended.
- Complete additional investigations if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.

**Emergency Department Monitoring:**

- Obtain baseline vital signs.
- Monitor temperature, heart rate, respiratory rate, blood pressure, and oxygen saturation.
- Monitor hydration status and fluid intake and output.
- Observe closely for deterioration in clinical status.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (APVIC) guidelines for management of fever.

**Inpatient Care:**

- Administer analgesics as needed.
- In euvolemic patients with SCD who are unable to drink fluids provide intravenous hydration at no more than 100 ml/kg/hour.
- Administer O2 as per clinical status.
- Ensure that patient is receiving maintenance fluids.
- Monitor vital signs.
- Refer to the inpatient sickle cell order set in Epic.
- Consult Pediatric Medicine for admission to the Children’s Hospital at Brampton Civic Hospital.
- If required, high risk patients may be transferred to the Satellite SCD Centre at Brampton Civic Hospital.

**Consider Admission to ICU:**

- Acute chest syndrome.
- Shock.
- Anaemia.
- Acute hydroxyurea administration.

**Exclusions:**

- Patients with systemic fungal infections.
- Patients with neutropenic fever.
- Patients with meningitis.
- Patients with sepsis.
- Outpatient management of febrile illness in infants and young children with sickle cell anaemia.
- Outpatient management of fever in sickle cell patients with fever managed with IV ceftriaxone followed by 3d of PO Ceclor (cefaclor).

**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](#)