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:**

1. Complete history, physical exam, and real-time additional investigations using ED Sickle Cell Fever order set to rule out:
   - Febrile illness unlikely to have a local source (e.g. sepsis, infection)
   - Suspected acute chest syndrome
   - Other peripheral crises

2. Check hemoglobin level, creatinine, electrolytes, (CBC, Differential, reticulocyte count, platelet count) if indicated.

3. Consult hematology fellow on call prior to discharge home from ED.

4. Most children with SCD present to the ED with fever and in stable clinical condition.

**Emergency Department Monitoring:**

- New onset severe pain
- Temperature 38°C or greater
- Fever and clinical suspicion for meningitis
- New onset clinical symptoms
- New onset chest pain
- Hypotension

**Formal Discharge:**

- Unless patient stable:
  - Inpatient admission for monitors
  - Close follow up with haematology

**Formal Follow Up:**

- Sickle Cell Clinic
- Local clinic
- Center at Brampton Civic Hospital

**Printable Version**

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