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 ß-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

Child with Sickle Cell Disease (SCD) presents to ED with oral or rectal temperature > 38.5ºC or axillary temperature (0.5ºC).

Emergency Department Initial Assessment and Management:
- Complete history, physical exam, and multi-investigations using ED Sickle Cell with Fever order set.
- Start antibiotics within 60 minutes of presentation. If infection site is unclear, start antibiotics and follow-up with Infectious Disease consultation.
- Additional investigations: chest x-ray; abdominal ultrasound; complete blood count, differential, reticulocyte count; blood culture; urinalysis.
- Serum electrolytes (sodium, potassium, creatinine).
- CBC, differential, reticulocyte count.
- Coagulation studies. Consider neurominidase inhibitor as per protocol.
- If patient is neutropenic, consult Hematology service for guidance related to the management of Hydroxyurea.
- If patient is hypotensive or unstable, consult Pediatrics.
- ED discharge planning.
- Child discharged home from inpatient unit with appropriate follow-up if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome.

Emergency Department Follow-up Monitoring:
- Temperature ≥ 38.5ºC
- Cyanosis
- Tachypnea
- Wheezing
- Excessive diaphoresis
- Gastrointestinal symptoms
- Seizures
- Changes in level of consciousness
- Changes in mental status
- Hypotension
- Changes in vital signs
- Changes in physical examination

End excluding other potential diagnoses.

Refer to Inpatient Care for admission to SickKids at any time

Guideline has been developed to guide the practice of clinicians at the Hospital for Sick Children. Use of this guideline in any setting must be subject to the clinical judgment of those responsible for providing care. SickKids does not accept responsibility for the application of this guideline outside SickKids.
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
Fever: Guidelines for Management in Children with Sickle Cell Disease

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