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 influenzae 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 routine lab investigations according to the SickKids Sickle Cell Management Document.
2. Completeness of physical exam is essential to determine signs of infection.
3. Blood culture in all patients with sickle cell disease and fever.
4. Pediatrician to review patient's medical record to determine if patient is admitted to the ED.
5. Management of fever for sickle cell disease as per hospital policy.
6. Management of chest pain, hypoxia or clinical suspicion for pneumonia or acute chest syndrome.
7. Education and support of the patient and family.
8. Follow-up with pediatrician.

**Emergency Department Monitoring:**
- Monitor heart rate, respiratory rate, and saturation.
- Administer antipyretics.
- Administer analgesics as needed.
- Complete history and physical exam.
- Monitor vital signs.
- Complete physical exam to include baseline pulse oximetry reading.
- History to include social history, vaccination history, surgical history, previous admissions.
- Current medications.
- Baseline pulse oximetry reading.
- Complications of SCD.
- Use of hydroxyurea.
- Use of folic acid.
- Use of penicillin prophylaxis.
- Use of other prophylactic agents.
- Development of chest crisis.
- Use of ambulatory ventilator.
- Development of splenic sequestration.
- Use of occlusive crisis.
- Use of priapism.

**Physicians to be consulted:**
- Emergency Department Initial Assessment and Management.
- Hematology.
- Infectious Disease.
- Pediatric Medicine.
- Physical exam to include:
  - Baseline pulse oximetry reading.
  - Complications of SCD.
  - Use of hydroxyurea.
  - Use of folic acid.
  - Use of penicillin prophylaxis.
  - Use of other prophylactic agents.
  - Development of chest crisis.
  - Use of ambulatory ventilator.
  - Development of splenic sequestration.
  - Use of occlusive crisis.
  - Use of priapism.

**Additional investigations:**
- CBC, differential, reticulocyte count, platelet count.
- Sodium, potassium, creatinine, uric acid, alkaline phosphatase, AST, ALT, bilirubin, AP, CRP, RDW.
- Blood cultures.
- Chest x-ray.
- Urine dipstick and culture.
- C-reactive protein.
- Radiology.
- Influenza testing.
- Outpatient management of febrile illness in infants and young children with sickle cell anaemia. J Pediatr. 1990;117(5):736–39 [a retrospective review of sickle cell patients with fever managed with IV ceftriaxone followed by 3d of PO Cefcror (cefaclor)].

**Patient Education:**
- Fever:
  - Guidelines for Management in Children with Sickle Cell Disease
  - Fever: Guidelines for Management in Children with Sickle Cell Disease

**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