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 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 neurologic exam. Include the following:
  - Fever (oral or rectal temperature > 38.5ºC or axillary temperature > 37.5ºC)
  - Edema
  - Respiratory distress
  - Evidence of splenic sequestration
  - Neurologic changes

- Urinalysis and culture (if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome – obtain if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome)

- Pediatrics, Emergency, and Hematology consult as needed if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome

**Emergency Department Monitoring:**

- CBC, differential, reticulocyte count, platelet count, coagulation panel, liver and renal function testing

- If patient is neutropenic or admitted to the hospital, administer parenteral antibiotics. Refer to Acute Painful Episodes Vaso-Occlusive Crisis Formulary for consultation.

- CVP or PA catheter insertion

**Inpatient Care:**

- Increase intake to maintain euvelema

- Monitor and notify MD if saturation falls below 95%

- Monitor for deterioration in clinical status

- If patient requires supplemental oxygen, change to continuous oxygen

- Bone marrow aspiration may be performed if patient presents with chest pain or clinical suspicion for osteomyelitis

- Consult Hematology for guidance related to the management of Hydroxyurea

**Outpatient Management:**

- Refer to Outpatient Management of Fever in Children with Sickle Cell Disease

**Follow up:**

- Consult Hematology for guidance related to the management of Hydroxyurea

**Additional Investigations:**

- Infectious disease screening (e.g., chest X-ray, echocardiogram)

**Support Services:**

- Social work

**Information During Hospital Transfer:**

- All patients at high risk for readmission must be referred for CCAC for follow up.

**4.0 References**


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### 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