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:**

1. Complete history, physical exam, and additional investigations as per ED Sickle Cell Fever order set as per SickKids inpatient management document.
2. Obtain additional workup as per Sickle Cell Fever inpatient management document.
3. Order a complete blood count (CBC) with differential and reticulocyte count.
4. Administer antipyretics, oral or IV in children who have fever.
5. Consult Hematology for guidance related to the management of Hydroxyurea.
6. Consult Pediatric Medicine for admission to inpatient unit if high risk.
7. Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
8. Obtain Sickle Cell Fever order set as per SickKids inpatient management document.
9. Complete history and routine lab investigations.
10. Refer to Acute Painful Episodes document (in Epic).

**Investigations**

- Routine lab investigations
- Additional investigations

**Emergency Department Monitoring**

- Monitor vital signs
- In euvolemic patients with SCD who are unable to drink fluids, monitor fluid intake and output.
- If patient is neutropenic, monitor white blood cell count.
- Note: Consult Hematology for guidance related to the management of Hydroxyurea.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.

**Inpatient Care**

- Inpatient Care
- Consult Hematology for guidance related to the management of Hydroxyurea.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).
- Consult Pediatric Medicine for admission to inpatient unit if high risk.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).

**Guidelines**

- For inpatient management document
- For outpatient management document
- Consult Hematology for guidance related to the management of Hydroxyurea.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).
- Consult Pediatric Medicine for admission to inpatient unit if high risk.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).

**Adverse Events**

- Monitor for adverse events
- Consult Hematology for guidance related to the management of Hydroxyurea.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).
- Consult Pediatric Medicine for admission to inpatient unit if high risk.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).

**Discharge Planning**

- Discharge home from ED or inpatient unit with appropriate follow-up
- Consult Hematology for guidance related to the management of Hydroxyurea.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).
- Consult Pediatric Medicine for admission to inpatient unit if high risk.
- Refer to Acute Painful Episodes Vasoocclusive Crisis (POE VPC) guidelines.
- Refer to Acute Painful Episodes document (in Epic).

### 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](#)