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 non-radiological investigations using ED Sickle Cell Fever order set. 
2. Consider additional investigations as needed (e.g.: chest x-ray, abdominal ultrasound, etc.). 
3. Provide dose of oral antibiotics for fever (see below). 
4. Consider admission to inpatient setting if high risk criteria met or suspected acute chest syndrome.
5. Perform chest x-ray if suspected pneumonia.
6. Administer intravenous hydration if oral intake is inadequate and/or patient is dehydrated.
7. Administer antipyretics if temperature is >38.5°C in children or >37.5°C in infants.
8. Refer to Hematology service for all patients with the following: bone pain, fever, hypoxia, new or worsening organ dysfunction, change in mental status, hospitalization, severe organ failure, severe bacterial infection, documented bacterimia, presence of new infection, or clinical suspicion of infection.
9. Consult Pediatric Medicine for admission to inpatient setting if any high risk criteria met.

Emergency Department Monitoring:
- Complete vital signs
- Baseline pulse oximetry reading
- Baseline chest x-ray
- Inpatient admission may be recommended if high risk criteria met

Inpatient Care:
- Antipyretics may be ordered in the Epic system
- Consult Hematology service for all patients with the following: bone pain, fever, hypoxia, new or worsening organ dysfunction, change in mental status, hospitalization, severe organ failure, severe bacterial infection, documented bacterimia, presence of new infection, or clinical suspicion of infection
- Administer intravenous hydration if oral intake is inadequate and/or patient is dehydrated
- Administer antipyretics if temperature is >38.5°C in children or >37.5°C in infants
- Follow up in clinic

Investigations:
- Routine lab investigations
- Additional investigations
- CBC, differential, reticulocyte count, serum creatinine, evidence of infection
- Obtain if child presents with cough, chest pain, hypoxia or clinical suspicion for pneumonia/acute chest syndrome

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 outpatient follow up.pdf
- SCD pain plan_july 2015.pdf