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 consultation with hematology if patient has SCD.
2. Suspected pneumonia or acute chest syndrome: obtain chest x-ray and blood culture.
3. Assess for other complications:
   a. Stroke/TIA
   b. Hepatic failure
   c. Diabetic ketoacidosis
4. Inpatient management document:
   a. Tolerating fluids and medications by mouth
   b. Follow up on hospital admission

Emergency Department Monitoring:
1. Monitoring sites:
   a. Baseline temperature
   b. Oxygen saturation
   c. Vital signs
2. Evidence of focal infection:
   a. Clinical symptoms of urinary tract infection (UTI) or known renal anomaly
   b. Evidence of meningitis or septicemia
3. General appearance:
   a. Presence of jaundice or pallor
   b. Vital signs

Initial Care:
1. Fever management:
   a. Tylenol and Ibuprofen
2. Tolerating fluids and medications by mouth
3. Follow up on hospital admission

Discharge planning process:
1. Child to be discharged home from ED if:
   a. Child is tolerating fluids and medications by mouth
   b. Temperature < 38.5ºC
   c. Oxygen saturation > 95%
   d. Tolerating fluids and medications by mouth
2. Contact hematology for guidance related to the management of Hydroxyurea

Environmental Training:
1. General information:
   a. Infections
   b. Anemia
   c. Stroke/TIA
   d. Meningitis
2. Inpatient management:
   a. Tolerating fluids and medications by mouth
   b. Follow up on hospital admission

Routine lab investigations:
- CBC, differential, reticulocyte count, platelet count, neutral counts.
- Urine dipstick and culture.

Additional investigations:
- Chest x-ray if suspected and if treatment with parenteral antibiotics should be given even if there is an obvious focus of infection.
- Bone marrow aspirate if concerns about the patient’s clinical status.
- CBC, differential, reticulocyte count, platelet count, neutral counts if there is clinical suspicion for occult sepsis.

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