Fever: Guidelines for Management in Children with Sickle Cell Disease

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 hypo function. 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:
2. Repeat complete blood count (CBC) and white blood cell differential when indicated.
3. Administer antipyretics per 3.0.1
4. Refer to Emergency Department Monitoring if patient appears unwell, has suspected infection, or is unable to drink fluids.
5. Consider antibiotics in children with previous documented bacteremia.
6. Additional investigations as per 3.0.5.

Emergency Department Monitoring:
1. Monitor vital signs: heart rate, systolic blood pressure, temperature, respiration rate, oxygen saturation.
2. Monitor fluid intake and output.
5. Refer to Emergency Department Monitoring

Child with Sickle Cell Disease (SCD) presents to ED with fever (oral or rectal temperature > 38.5ºC or axillary temperature > 37.5ºC).

Investigations:
- Routine lab investigations
- Additional investigations
- CBC, differential, reticulocyte count, CO2, glucose, creatinine, sodium, potassium, calcium, magnesium, liver function tests, urinalysis, chest x-ray, blood culture (if suspected), influenza A/B and RSV test
- Follow up confirmed by telephone call or in person
- Inpatient admission
- Outpatient follow up
- Consultation with Hematology

Physiological stability:
- Complete lab investigations
- Complete history to include:
  - Children with SCD presenting with fever should be assessed for SCD complications:
    - Fever: Guidelines for Management in Children with Sickle Cell Disease
  - Complications of SCD
    - Acute splenic sequestration
    - Aplastic crisis
    - Acute chest syndrome
    - Appendicitis
    - Stroke
    - Osteomyelitis
    - Gallbladder disease
    - Septic arthritis
  - Evidence of focal infection
  - Transfusion history
  - Respiratory
    - Evidence of upper respiratory tract infection
    - Evidence of lower respiratory tract infection
  - Other medical history
    - History of previous admissions for VOC or readmissions for ACS
  - Allergies
  - Medications
    - Recent and current prescription medications
    - Significant urinary tract infections
  - Vaccination history
  - Baseline hemoglobin
  - Duration of fever
  - History of fever
  - History of history of fever
  - Duration of fever
  - History of history of fever
  - Duration of fever

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](Fever Care Pathway Final 2021.pdf)
- [Revision History.docx](Revision History.docx)
- [SC_Clinic Follow Up Revised 2021_FINAL.pdf](SC_Clinic Follow Up Revised 2021_FINAL.pdf)
- [SCD fever_criteria for admission 2021 FINAL.pdf](SCD fever_criteria for admission 2021 FINAL.pdf)
- [SCD fever_discharge planning process 2021 FiNAL.pdf](SCD fever_discharge planning process 2021 FiNAL.pdf)
- [SCD fever_inpatient management.pdf](SCD fever_inpatient management.pdf)
- [SCD fever_out patient follow up.pdf](SCD fever_out patient follow up.pdf)
- [SCD pain plan_july 2015.pdf](SCD pain plan_july 2015.pdf)