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:
1. Complete history, physical exam, and multi-organ additional investigations using ED Sickle Cell Fever order set.
2. Conduction of blood culture if suspected: a) pneumococcal pneumonia, b) sepsis, c) pneumonia, d) meningitis.
3. Place patient on IV fluids as per Age Group/Upper limit of normal, infusion rate to be determined by paediatrician.
4. Place patient on oxygen if hypoxia.

Inpatient Care
1. Child with SCD who presents to ED with fever and/or rectal temperature > 38.5ºC or axillary temperature 38ºC should be admitted.
2. Provide dose of oral antibiotics for treatment of SCD.
3. Give IV ceftriaxone followed by 3 days of PO Ceclor (cefaclor).
4. Monitor vital signs.
5. Consult Haematology service for all patients with the following clinical symptoms: 
   - Macrocytic anaemia, 
   - Acute chest syndrome (ACS) – pneumococcal pneumonia, 
   - Pneumonia/acute chest syndrome – obtain if child presents with cough, chest pain, hypoxia or clinical suspicion for.
6. Ensure that patient is receiving maintenance fluids.
7. Baseline hemoglobin, baseline pulse oximetry reading.
8. Baseline sodium, potassium.
9. CBC, differential, reticulocyte count.
10. Document laboratory results.

Inpatient Management
1. Child has stable vital signs including blood pressure and heart rate.
2. Tolerating fluids and medications by mouth.
4. Follow up and discharge.

Infantile Dose
1. Child with SCD who presents to ED with fever and oral temperature > 38.5ºC or axillary temperature 38ºC should be admitted.
2. Provide dose of oral antibiotics for treatment of SCD.
3. Give IV ceftriaxone followed by 3 days of PO Ceclor (cefaclor).
4. Monitor vital signs.
5. Consult Haematology service for all patients with the following clinical symptoms:
   - Macrocytic anaemia, 
   - Acute chest syndrome (ACS) – pneumococcal pneumonia, 
   - Pneumonia/acute chest syndrome – obtain if child presents with cough, chest pain, hypoxia or clinical suspicion for.
6. Ensure that patient is receiving maintenance fluids.
7. Baseline hemoglobin, baseline pulse oximetry reading.
8. Baseline sodium, potassium.
9. CBC, differential, reticulocyte count.
10. Document laboratory results.

Inpatient Management
1. Child has stable vital signs including blood pressure and heart rate.
2. Tolerating fluids and medications by mouth.
4. Follow up and discharge.

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