

## Fever: Guidelines for Management in Children with Sickle Cell Disease

Version: 6

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### 1.0 Introduction

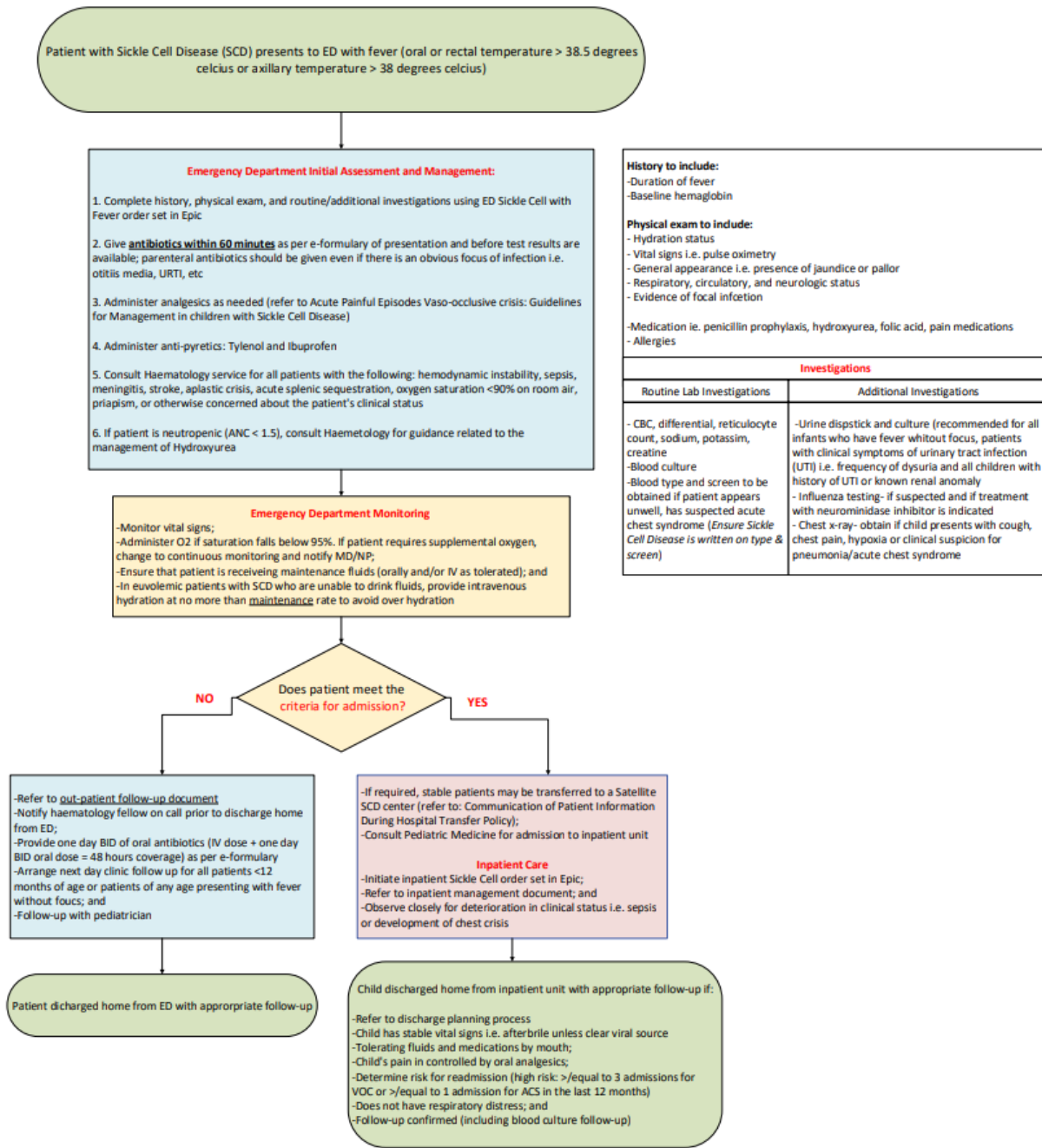
By 3–4 months of age (when fetal hemoglobin declines to <50% of total), many children with sickle cell anemia (HbSS) and sickle  $\beta$ -thalassemia develop clinically significant hemolytic anemia and impairment of splenic function. In others, although the HbF may remain above 50% these children are 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 pneumoniae* and *Haemophilus influenzae* 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 influenzae 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  $>38.5^{\circ}\text{C}$  or an axillary temperature  $>38^{\circ}\text{C}$ , 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



### Copy of Printable Flow Chart

### 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](#)