

Fever: Guidelines for Management in Children with Sickle Cell Disease

Version: 5

This is a CONTROLLED document for internal use only, valid only if accessed from the Policies and Procedures site.

1.0 Introduction

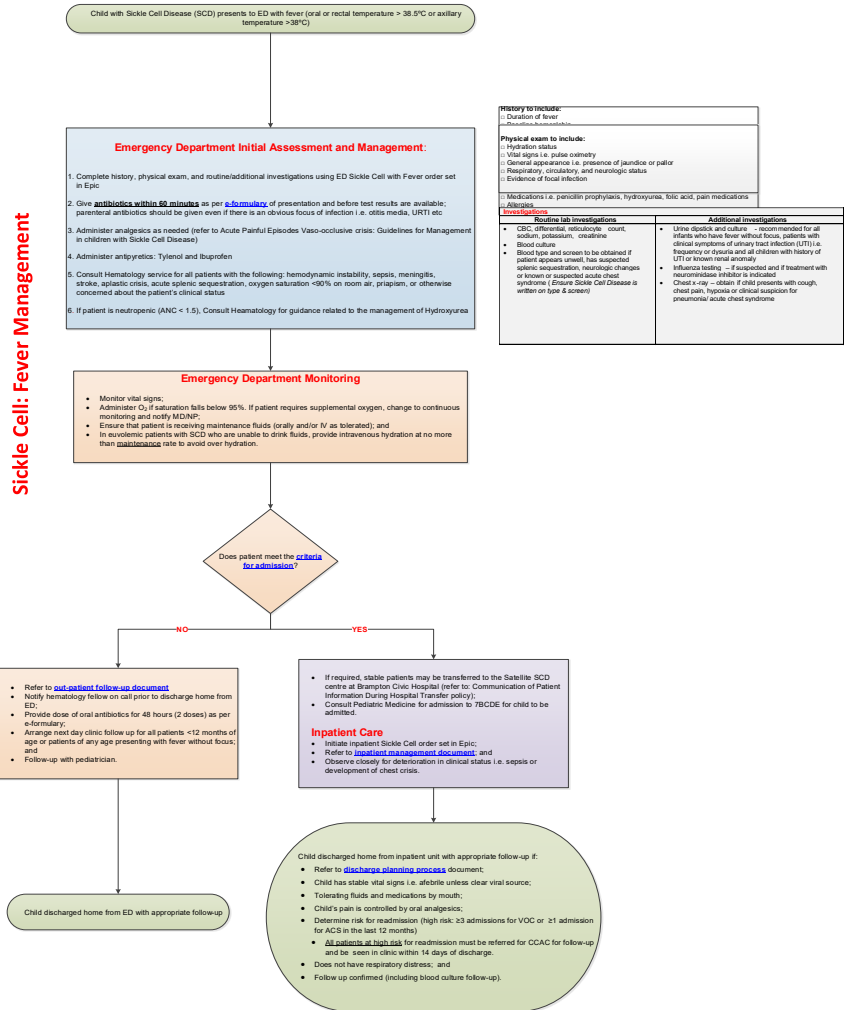
By 3–4 months of age (when fetal hemoglobin declines to <50% of total), many children with sickle cell anemia (HbSS) and sickle β -thalassemia develop clinically significant hemolytic anemia and impairment of splenic function. In others, although the HbF may remain above 50% these children are still 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 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



PRINTABLE VERSION

4.0 References

1. Chesney PJ, Wilimas JA, Presbury G, et al. Penicillin and cephalosporin-resistant strains of Streptococcus pneumoniae causing sepsis and meningitis in children with sickle cell disease. *J Pediatr.* 1995;127(4):526–32.
2. Rogers ZR, Morrison RA, Vedro DA, et al. Outpatient management of febrile illness in infants and young children with sickle cell anaemia. *J Pediatr.* 1990;117(5):736–39 [a retrospective review of sickle cell patients with fever managed with IV ceftriaxone followed by 3d of PO Ceclor (cefaclor)].

©The Hospital for Sick Children ("SickKids"). All Rights Reserved. This document was developed solely for use at SickKids. SickKids accepts no responsibility for use of this material by any person or organization not associated with SickKids. A printed copy of this document may not reflect the current, electronic version on the SickKids Intranet. Use of this document in any setting must be subject to the professional judgment of the user. No part of the document should be used for publication without prior written consent of SickKids.

3. Wilimas JA, Flynn PM, Harris SC, et al. A randomized study of outpatient treatment with ceftriaxone for selected febrile children with sickle cell disease. *N Engl J Med*. 1993;329(7):472–76 [a randomized study of children with sickle cell disease and fever at low risk for sepsis treated with two doses of IV ceftriaxone q24h vs. inpatient management].
4. Williams LL, Wilimas JA, Harris SC, et al. Outpatient therapy with ceftriaxone and oral cefixime for selected febrile children with sickle cell disease. *J Pediatr Hematol Oncol*. 1996;18(3):257-61 [a prospective, nonrandomized study on use of IV ceftriaxone followed by PO cefixime for febrile sickle cell patients at low risk for sepsis]
5. Peter G. (ed): *1997 Red Book: Report of the Committee on Infectious Diseases*. 24th ed. American Academy of Pediatrics.
6. Savlov, D., Beck, C.E., DeGroot, J., Odame, I., & Friedman, J.N. (2014). Predictors of bacteremia among children with sickle cell disease presenting with fever. *Journal of Pediatric Hematology Oncology*, 36(5), 384-388.
7. West, D.C., Andrada, E., Azari, R., Rangaswami, A.A, & Kuppermann, N. (2002). Predictors of bacteremia in febrile children with sickle cell disease. *Journal of Pediatric Hematology Oncology*, 24(4), 279-283.
8. Chang, T.P., Kriengsoontorkij, W., Chan, L.S., & Wang, V.J. (2013). Predictors for bacteremia in febrile sickle cell disease children in the post-7-valent pneumococcal conjugate vaccine era. *Journal of Pediatric Hematology Oncology*, 35 (5), 377-382.
9. Olivares, M., Richardson, M.W. & Visintainer, P. (2013). C-reactive protein as a predictor of bacterial disease in children with sickle cell disease and fever. *Blood*,122(21).
10. Baskin, M.N., Goh, X.L., Heeney, M.M., Harper, M.B. (2013). Bacteremia risk and outpatient management of febrile patients with sickle cell disease, *Pediatrics*,131(6), 1035-1041.
11. National Heart, Lung and Blood Institute (2014). Evidence based management of sickle cell disease: Expert panel report, Available at: <https://www.nhlbi.nih.gov/health-topics/evidence-based-management-sickle-cell-disease>

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](#)