Table 4: Evidence Supporting Appropriate Emergency Department Fever Management for Children with Sickle Cell Disease

Type of evidence	Key findings	Level of evidence (USPSTF ranking*)	Citation(s)
Clinical guidelines	All children with SCD who have fever greater than 38.5 degrees Celsius or 101 degrees Fahrenheit and other signs of infection should be evaluated promptly. The younger the child, the higher the index of suspicion. In a child with no obvious sources of infection, a minimum evaluation should include blood culture, complete blood count, reticulocyte count, and chest x-rays for children under 3 years of age. Immediately after the blood is taken, the child should be given broad-spectrum antibiotics, preferably intravenously. Broad spectrum antibiotics should be given even if these tests cannot be performed (p. 28). Ideally, children with SCD are followed at a practice or center that allows for comprehensive management of their disease. These facilities should have 24-hour access to medical consultants, hematology and microbiology laboratories, and a blood bank, among other services (p. 29).	III	National Heart Lung and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health. Bethesda, MD, 2002.
Clinical guidelines	A child with fever or pallor and listlessness should always be initially evaluated, if possible, at a site where complete blood cell (CBC) and reticulocyte counts, blood cultures, intravenous antibiotics, and red blood cell transfusions are readily available. Because patients with SCD develop splenic dysfunction at as early as 3 months of age, they are at high risk for septicemia and meningitis with pneumococci and other encapsulated bacteria. Thus, all patients with temperature greater than 38.5 degrees C require rapid triage and physical assessment, urgent CBC and reticulocyte counts, blood culture (plus cerebrospinal fluid analysis and other cultures as indicated), and prompt administration of a broad-spectrum parenteral antibiotic, such as ceftriaxone sodium, cefuroxime, or cefotaxime sodium (p. 529).	III	American Academy of Pediatrics Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. *Pediatrics*. Mar 2002;109(3):526-535.
Clinical guidelines	Children with SCD with a temperature greater than 38.5 degrees C should be promptly administered IV ceftriaxone (50-100 mg/kg, 2.0 maximum dose. Relatively high doses (75-100 mg/kg) are sometimes recommended in regions with high prevalence of antibiotic resistant S.	III	Lane PA, Buchanan GR, Hutter JJ, et al. Sickle cell disease in children and adolescents: diagnosis, guidelines for comprehensive care,

Type of evidence	Key findings	Level of evidence (USPSTF ranking*)	Citation(s)
Clinical guidelines (continued)	pneumoniae. • Strongly consider adding vancomycin (10-15 mg/kg IV) for severe illness or if CNS infection is suspected. • Parenteral antibiotics should be given before other procedures. • The presence of a focus of infection does not alter the urgency of giving parenteral antibiotics (p. 13).	III	and care paths and protocols for management of acute and chronic complications. 2001; Annual Meeting of the Sickle Cell Disease Care Consortium, Sedona, AZ
Clinical guidelines	Children with SCD with a fever greater than or equal to 38.5 degrees C should be given parenteral broad spectrum antibiotic treatment within 60 minutes of triage	III	Wang CJ et al. Quality-of-care indicators for children with sickle cell disease. *Pediatrics* 2011; 128:484-493.

Note: USPSTF criteria for assessing evidence at the individual study level are as follows: I) Properly powered and conducted randomized controlled trial (RCT); well-conducted systematic review or meta-analysis of homogeneous RCTs. II) Well-designed cohort or case-control analytic study. III) Opinions of respected authorities, based on clinical experience; descriptive studies or case reports; reports of expert committees.