Table 5: Evidence Supporting Emergency Department Pain Assessment for Children with Sickle Cell Disease

Type of Evidence	Key Findings	Level of Evidence (USPSTF Ranking*)	Citation(s)
Clinical guidelines	The NHLBI suggests the following for children with SCD who are experiencing pain: Education about pain management is the basis for collaboration among patients, families, and health care providers for optimal treatment. Patients must be reassured that when they do experience pain it will be taken seriously and managed optimally with a plan. Because patient needs change over time, plans for their care must be assessed and modified accordingly. Clinicians should ask about pain and use patients' reports as the primary source for assessment, except in infants where behavioral observations are the main basis for evaluation. Most SCD pain can be managed well if the barriers to assessment and treatment are overcome; a comprehensive psychosocial clinical assessment should be performed yearly (more often for patients with frequent pain). SCD pain can be described as: • Acute – The most common type of SCD pain characterized by an unpredictably abrupt onset without any other explanation. Intensity varies from mild to severe and can last from hours to a few days. Pain may reoccur and migrate from one site to another. • Chronic – Pain that lasts 3 to 6 months or more and no longer serves a warning function. The condition may be hard to distinguish from frequently recurring acute pain and can be debilitating both physically and psychologically. • Mixed – Pain frequently is mixed as to type and mechanism. Clinicians should understand the pain in detail to tailor therapy to the needs of the patient. Assessment depends on the chronologic age, developmental stage, functional status, cognitive ability, and emotional state, so these factors should be considered in the choice of measurement tools. Frequent reassessment is important. Pain management should be aggressive to relieve pain and achieve maximum function. Analgesics are the foundation for the management of sickle cell pain, and their use should be tailored to the individual patient. Pharmacological pain management consists of the use of nonsteroidal anti-inflammatory drugs (NSAIDs), opioids, and adjuvant medications. • Management of mild-to-moderate pain should include NSAIDs or acetaminophen • If mild-to-moderate pain persists, and opioid can be added. • Persistent or moderate-to-severe pain relies on repeated assessments and appropriate increases in opioid strength or dose.	III	National Heart Lung and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health. Bethesda, MD, 2002.

Type of Evidence	Key Findings	Level of Evidence (USPSTF Ranking*)	Citation(s)
Clinical guidelines	The AAP sections on Hematology/Oncology and the Committee on Genetics suggest the following for children with SCD experiencing pain: Recognition and appropriate management of painful events should be reviewed as part of family/patient education during regularly scheduled visits. The ultimate goal is to enable families to functionally cope with a child's complex chronic illness and transition successful to adulthood. Many uncomplicated episodes can be managed at home with oral fluids; oral analgesics (ibuprofen, acetaminophen) and codeine; and comfort measures, such as heating pads. For severe pain, parenteral opioids (e.g., morphine) are administered, usually with around-the-clock dosing or patient-controlled analgesia. Opioids should not be withheld because of the unfounded fear of addiction. Health care providers should maintain patients on adequate, but not excessive hydration; oxygen and cardio-pulmonary status should be monitored; and patients should be watched for other developments like acute chest syndrome.	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	Patients with an uncomplicated vaso-occlusive pain event may have few physical symptoms to suggest the severity of their pain on presentation. Pain assessment relies on patient self-report; pain intensity may be assessed using several available scales including the visual analog scale, verbal scale, and Wong-Baker face scale for children. The choice of tool will depend on the age, cognitive ability, and emotional state. Pain scores should be documented and used to modify the treatment plan, as needed.	III	Ellison AM, Shaw K. Management of vaso- occlusive pain events in sickle cell disease. Pediatr Emerg Care 2007; 23(11):832-838.
Clinical guidelines	Effective pain management requires frequent assessment to maintain pain control, make adjustments due to tolerance or adverse effects of opioid therapy, and identify exacerbations of pain and/or other complications. Once a thorough assessment has been completed, a comprehensive management approach, including appropriate pharmacological, psychological, behavioral, and physical strategies, can be implemented. A comprehensive, multifaceted pain assessment is designed to individualize care and examine the overall needs of the child and family. A simple measure of pain intensity that can be quickly completed, accounting for the child's development stage and cognitive abilities, is essential. Pain intensity should be assessed initially, after the peak effect of the medication, and at frequent intervals until adequacy and duration of the medication's effects have been determined.	III	Stinson J, Naser B. Pain management in children with sickle cell disease. Pediatr Drugs 2003; 5(4):229-241.

Type of Evidence	Key Findings	Level of Evidence (USPSTF Ranking*)	Citation(s)
Clinical guidelines	The optimal treatment of acute sickle cell vaso-occlusive pain requires experienced clinicians providing rapid evaluation and aggressive treatment with supportive care and analgesics. In the absence of such expertise, a clinical practice guideline may provide the framework for appropriate assessment and treatment. Educating health care providers about behaviors often misinterpreted as addiction and about the very low true rates addiction, combined with the use of a clinical practice guideline, are significant steps in influencing prescribing practices in SCD patients.	III	Morrissey LK, O'Brien, Shea J, Kalish LA, Weiner DL, Branowicki P, Heeney MM. Clinical practice guideline improves the treatment of sickle cell disease vaso-occlusive pain. Pediatr Blood Cancer 2009; 52:369-372.
Clinical guidelines	There's no standard method for treating pain. One approach consists of the following steps: treat the cause, if possible; begin analgesics; start fluids; for acute pain, administer an opioid; for chronic pain, use fentanyl patches, acetaminophen, codeine and NSAIDs. Patients with severe pain should be given an opioid parenterally at frequent, fixed intervals, not as needed, until the pain has diminished, when the opiate dose can be tapered and oral analgesics started. Management of constant pain is extremely difficult; expert advice should be obtained. Most patients with acute pain are neither drug addicts nor seekers. Reliable patients can be given oral analgesics with codeine at home.	III	Steinberg MH. Management of sickle cell disease. N Engl J Med 1999; 340(13):1021-1030.
Clinical guidelines	Pain management for children with SCD should be consistent, aggressive, and tailored to meet individual needs, acute or chronic. Pharmacological management for SCD pain may include NSAIDs, opioids, and adjuvant medications. For mild-to-moderate cases, NSAIDs, acetaminophen, and tramadol are useful; for moderate-to-severe pain, opioids (codeine, hydrocodone, etc.) should be used. Primary care physicians should consult with a hematologist or SCD specialist on the management of moderate-to-severe pain.	III	Pack-Mabien A, Haynes Jr J. A primary care provider's guide to preventive and acute care management of adults and children with sickle cell disease. J Am Acad Nurse Pract 2009; 21:25-257.

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.