

Table 4: Evidence Supporting Anticipatory Guidance of the Prevention and/or Management of Pain in Children with Sickle Cell Disease

| TYPE OF EVIDENCE | KEY FINDINGS | LEVEL OF EVIDENCE (USPSTF RANKING*) | CITATION(S) |
|-----------------------------------|--|-------------------------------------|---|
| <p>Clinical guidelines</p> | <p>The NHLBI suggests the following for children with SCD who are experiencing pain:</p> <p>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).</p> <p>SCD pain can be described as:</p> <ul style="list-style-type: none"> • 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. <p>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.</p> <p>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.</p> | <p>III</p> | <p>National Heart Lung and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health. Bethesda, MD, 2002.</p> |

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| | <ul style="list-style-type: none"> • 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. | | |
| Clinical guidelines | <p>The AAP sections on Hematology/Oncology and the Committee on Genetics suggest the following for children with SCD experiencing pain:</p> <p>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.</p> <p>Many uncomplicated episodes can be managed at home with oral fluids; oral analgesics (ibuprofen, acetaminophen) and codeine; and comfort measures, such as heating pads.</p> <p>For severe pain, parenteral opioids (e.g., morphine) are administered, usually with around-the-clock dosing or patient-controlled analgesia.</p> <p>Opioids should not be withheld because of the unfounded fear of addiction.</p> <p>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.</p> | III | American Academy of Pediatrics Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. <i>Pediatrics</i> . Mar 2002;109(3):526-535. |
| Clinical guidelines | <p>Few advances have been made in the acute management of vaso-occlusive pain episodes. Successful therapy relies on thorough assessment and reassessment; aggressive analgesia; adequate hydration; and close monitoring for the development of other disease complications.</p> <p>Treatment of pain may include the following steps: assessment of the vaso-occlusive episode; pharmacological management; adjuvant medications; non-pharmacological interventions; blood transfusions/ physiotherapy/oxygen therapy/ hydration/hydroxyurea.</p> <p>Behavioral, cognitive, and physical therapies should be part of a comprehensive approach. These may include distraction, guided imagery, hypnosis, breathing exercises, heat, cold, and massage therapy.</p> | III | Ellison AM, Shaw K. Management of vaso-occlusive pain events in sickle cell disease. <i>Pediatr Emerg Care</i> 2007; 23(11):832-838. |

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| Clinical guidelines | <p>For SCD pain, acetaminophen and ibuprofen alone have little efficacy, so are best when given in combination with more potent medications, for mild or brief pain, or in situations such as work or school, when sedation should be avoided.</p> <p>Time-contingent schedules are important, especially for patients with severe pain. Analgesic combinations with potent opioids are more effective than single medications. Use of longer-acting medications to reduce the complexity of administration schedules is appropriate for patient dealing with severe pain.</p> | III | Dampier C, Ely E, Brodecki D, O'Neil P. Home management of pain in sickle cell disease: A daily diary study in children and adolescents. <i>J Pediatr Hematol Oncol</i> 2002; 24(8):643-647. |
| Clinical guidelines | <p>Pain can be effectively managed in most children by aggressive use of currently available treatment approaches. Effective management requires frequent assessment to maintain pain control, appropriate adjustments due to tolerance or adverse effects of opioid therapy, and identification of 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 to reduce pain, improve functional ability, and enhance well-being.</p> | III | Stinson J, Naser B. Pain management in children with sickle cell disease. <i>Pediatr Drugs</i> 2003; 5(4):229-241. |
| Clinical guidelines | <p>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.</p> <p>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.</p> <p>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.</p> | III | Steinberg MH. Management of sickle cell disease. <i>N Engl J Med</i> 1999; 340(13):1021-1030. |
| Clinical guidelines | <p>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</p> | 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. <i>J</i> |

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| | physicians should consult with a hematologist or SCD specialist on the management of moderate-to-severe pain. | | <i>Am Acad Nurse Pract</i> 2009; 21:25-257. |
| Clinical guidelines | <p>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.</p> <p>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.</p> | III | <p>Morrissey LK, O'Brien Shea J, Kalish LA, Weiner DL, Branowicki P, Heeney MM. Clinical practice guideline improves the treatment of sickle cell disease vasoocclusive pain. <i>Pediatr Blood Cancer</i> 2009; 52:369-372.</p> |

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.