Sickle Cell Anemia and Pain: Will Data Prevail Over Beliefs?

Richard Payne, MD
From the Institute on Care at the End of Life, Duke Divinity School, Durham, NC.

0196-0644/$-see front matter
Copyright © 2008 by the American College of Emergency Physicians.
doi:10.1016/j.annemergmed.2008.10.022

SEE RELATED ARTICLE, P. 587.

Pain is a cardinal feature of sickle cell anemia, and its treatment is a major clinical problem because pain associated with vaso-occlusive events is unpredictable in its timing, can be quite severe, and adversely affects quality of life. Optimal management of sickle cell–related pain requires expertise in assessment and skill in the use of potent parenterally administered opioid analgesics. Furthermore, painful episodes may signal life-threatening medical complications, such as sepsis, acute chest syndrome, stroke, blood and fat pulmonary embolism, and organ failure. In fact, an acute pain episode precedes 22% of sickle cell deaths.¹

Emergency departments (EDs) are by far the most common site for treatment of acute, severe, sickle cell–related pain, and this setting poses additional challenges to high-quality pain management. First of all, current crowded conditions in the nation’s EDs can overwhelm our capacity to treat pain in a timely fashion.² A recent study of sickle cell pain in 3 EDs documented a median time of 90 minutes to analgesic administration, with some patients waiting up to 10 hours for their initial dose.³ A lack of well-accepted emergency medicine standards for sickle cell treatment may contribute to delays in care. In fact, only 20% of teaching hospitals have protocols promoting rapid, efficient, and evidence-based management of sickle cell pain.⁴

Clinicians’ attitudes toward pain, addiction, and possibly patients themselves also influence sickle cell pain management. When physicians were surveyed anonymously at 7 National Institutes of Health–funded sickle cell centers and asked to rank their beliefs about barriers to optimal management of sickle cell pain, “fear that the patient is drug abuser,” “reluctance to prescribe opioids,” and “disbelief in patient’s report of pain severity” ranked in the top 5 of 15 possible respondent choices.⁵

In a similar study of nursing attitudes about sickle cell pain management, 63% of the 77 respondents believed that “drug addiction frequently develops in the treatment of sickle cell pain episodes,” and 30% were “hesitant to administer high-dose opioids,” although, paradoxically, the majority of respondents (86%) believed that “drug addiction should not be a primary nursing concern when caring for a patient with sickle cell pain episodes.” These studies echo a previous survey reporting that 53% of emergency physicians and 23% of hematologists believe that more than 20% of patients with sickle cell disease are addicted.⁷

These perceptions of pain and addiction must also be placed in a larger social context of sickle cell disease, which has been erroneously characterized as an exclusively “black disease” (in fact, it occurs primarily in people of African descent, but also in Mediterranean and other ethnic groups).⁸ However, the predominance of sickle cell anemia in black patients inevitably connects their pain experiences to the pervasive influence of racial and ethnic disparities within the health care system generally and pain management specifically.⁹ Many recent reports document that patients’ behaviors and communication styles may complicate pain assessment by reinforcing conscious and unconscious stereotyping, leading to a discounting of the patient’s pain complaints.¹⁰ Not uncommonly, sickle cell patients who require frequent admissions to the ED for pain complaints are labeled pejoratively as “frequent fliers” and “drug-seekers.” Emotionally loaded terms such as these surely affect the assessment of subjective clinical phenomena such as pain and have been shown to influence health care provider attitudes about minority patients.¹¹

It is in this context that the report of Aisiku et al¹² in this edition of Annals of Emergency Medicine is most appreciated and welcomed.¹² These investigators present new and long-awaited clinical information comparing 82 sickle cell patients who utilized EDs 3 or more times per year for treatment of pain (high utilizers) to 150 patients who used the ED fewer than 3 times per year (low utilizers). This important study confirms that high utilizers have greater disease severity, as evidenced by lower hematocrit levels and higher transfusion requirements than low utilizers. Furthermore, daily diaries record that high utilizers were more symptomatic, with higher mean pain and distress scores and greater negative effects of the disease and pain on Short Form-36 questions quality of life measures. After controlling for the number of pain days and the magnitude of pain, high utilizers did not use opioids more frequently than low utilizers, which suggests that disease severity and pain were responsible for opioid use, rather than opioid use being driven by compulsive behavioral consumption associated with psychological dependence. As Aisiku et al¹² remarked, however, frequent ED use does not preclude coexisting problems such as alcohol abuse and psychiatric dysfunction, but their data do strongly suggest that sickle cell patients are not unlike other
populations who require frequent ED care—they have clinically significant acute and chronic medical problems.\textsuperscript{13}

This study should dispel some myths about sickle cell patients who experience frequent pain and should correct misperceptions of health care providers about frequent ED utilizers. In addition, the management of sickle cell–related pain in EDs would be greatly improved by adoption of treatment protocols based on evidence-based guidelines. Guidelines such as those created by the American Pain Society emphasize the need for rapid attention to patients reporting severe pain.\textsuperscript{14}

Published ED guidelines recommend high triage levels (level 1 or 2) for patients with pain scores greater than 7 (on a 0 to 10 scale), and sickle cell pain protocols should be consistent with this recommendation.\textsuperscript{15} The American Pain Society guidelines also recommend intravenous or subcutaneous administration of appropriate doses of morphine or hydromorphone (rather than meperidine) within 20 minutes of admission to the ED. In general, more education of ED and other health care providers who treat sickle cell patients is needed to improve relevant skills and confidence in pain assessment and management, particularly in the dosing of opioid analgesics and the management of adverse effects.\textsuperscript{5}

Finally, alternatives to ED management of sickle cell patients should be considered. Benjamin et al\textsuperscript{16} reported on the use of a day-hospital facility, staffed with experts in sickle cell disease and pain management who have long-term relationships with their patients. Very encouragingly, they demonstrated that, when compared with general ED treatment, day-hospital care resulted in more rapid pain control, fewer ED visits for sickle cell–related pain, fewer admissions, and shorter lengths of stay for hospitalized patients. Finally, they noted a cost savings of approximately $1.7 million in a large teaching hospital,\textsuperscript{16} providing a financial incentive for replication of this model in other institutions.

Albert Schweitzer remarked that “pain is a more terrible lord of mankind than even death itself.” Congratulations to Aisiku et al\textsuperscript{12} for providing data to confront the unfortunate rhetoric and stereotyping that has handicapped the management of patients with sickle cell–related pain for far too long.

\textbf{Supervising editor:} Knox H. Todd, MD, MPH

\textbf{Funding and support:} By Annals policy, all authors are required to disclose any and all commercial, financial, and other relationships in any way related to the subject of this article that might create any potential conflict of interest. The author has stated that no such relationships exist. See the Manuscript Submission Agreement in this issue for examples of specific conflicts covered by this statement.

\textbf{Reprints not available from the author.}

\textbf{Address for correspondence:} Richard Payne, MD, Institute on Care at the End of Life, Duke Divinity School; 919-660-3553; E-mail rpayne@div.duke.edu.

\textbf{REFERENCES}


\textbf{Did you know?}

You can track the impact of your article with citation alerts that let you know when your article (or any article you’d like to track) has been cited by another Elsevier-published journal.

Visit [www.annemergmed.com](http://www.annemergmed.com) today to see what else is new online!