



Improving Emergency Department-Based Care of Sickle Cell Pain

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Pain is the leading cause of emergency department (ED) visits for individuals living with sickle cell disease (SCD). The care that is delivered in the ED is often cited by patients with SCD as the area of health care in greatest need of improvement. In 2014, the National Heart, Lung, and Blood Institute released guidelines for the care of SCD, including recommendations for the management of acute sickle cell pain in the ED. These guidelines provide a framework to understand the elements of ideal emergency sickle cell pain care; however, they do not provide guidance on barriers and facilitators to achieving these ideals in the complex system of the ED. Presented in this article are 4 tenets of implementing guideline-adherent emergency sickle cell care gleaned from the available literature and continuous quality improvement efforts at our institution. These include: (1) strategies to reduce negative provider attitudes toward patients with SCD; (2) strategies to reduce time-to-first-dose of analgesic medication; (3) strategies to improve ED pain care beyond the first dose of medication; and (4) strategies to improve ED patient safety. Application of the principles discussed within can improve patient and provider satisfaction, quality, and safety.

Learning Objectives

- Learn 2 complementary approaches to reduce negative emergency provider attitudes toward patients with sickle cell disease (SCD): provider education and intensive management of challenging patients
- Understand strategies to reduce time-to-first-dose of analgesic medication for patients with SCD who present to the ED with acute pain
- Understand strategies to safely increase the frequency of assessments and analgesic doses for ED patients with acute sickle cell pain

Introduction

Sickle cell disease (SCD) is a recessively inherited family of hemoglobin disorders that affects ~100 000 individuals in the United States and millions worldwide. In individuals living with SCD, deoxygenated hemoglobin forms rigid polymers that damage red blood cell membranes, activating various abnormal cell-signaling pathways and ultimately leading to the manifestations of the disease. Clinically, SCD is marked by hemolytic anemia, progressive organ damage, vaso-occlusion, and premature mortality, but the most salient clinical feature of the disease is pain. SCD pain accounts for the majority of health care costs related to SCD and is also the leading cause of emergency department (ED) visits and hospital admissions.¹ The care that is delivered in the ED is often cited by patients with SCD as the area of health care in greatest need of improvement. The present article reviews several strategies and the best available evidence for improving quality and patient experience for SCD pain care in the ED.

Sickle cell pain is complex and multifactorial. Vaso-occlusion, occurring primarily in postcapillary venules, is believed to be the primary etiology of acute, episodic sickle cell pain.² Pain can occur anywhere but most frequently involves bony areas where marrow is present, particularly the lower back and the legs, and in children the hands can be affected.³ Pain episodes often have a prodromal crescendo and resolution phase that lasts from a few days to several weeks.⁴

In adolescents and adults with SCD, many etiologies other than vaso-occlusion contribute to pain, including avascular necrosis, regional pain syndromes, neuropathic pain, opioid-induced hyperalgesia, and depression.⁵ For these reasons, most adults with SCD use prescription opioids at home, and 38% use long-acting opioids.⁶ Opioid tolerance is common and makes it very challenging to relieve or even reduce pain during acute exacerbations of the disease. Although subpopulations have higher rates of utilization, the average number of ED visits per year for acute pain is low, ranging from 1 to 7.5 depending on the population studied.^{7,8}

Very often, when individuals experience sickle cell pain, they have no option other than to seek treatment in a 911-receiving ED. EDs are complex systems, designed with limited resources to meet the needs of all patients experiencing medical emergencies. Several aspects of emergency care systems pose challenges to providing optimal care to individuals with SCD. Educational gaps and biases among providers, staff, and patients create barriers to communication and trust, and erode the provider-patient relationship. Scarcity of resources can jeopardize patient safety and limit access to needed treatments. Lack of institutional and departmental treatment protocols can lead to wide variability in the quality of care, resulting in patient and

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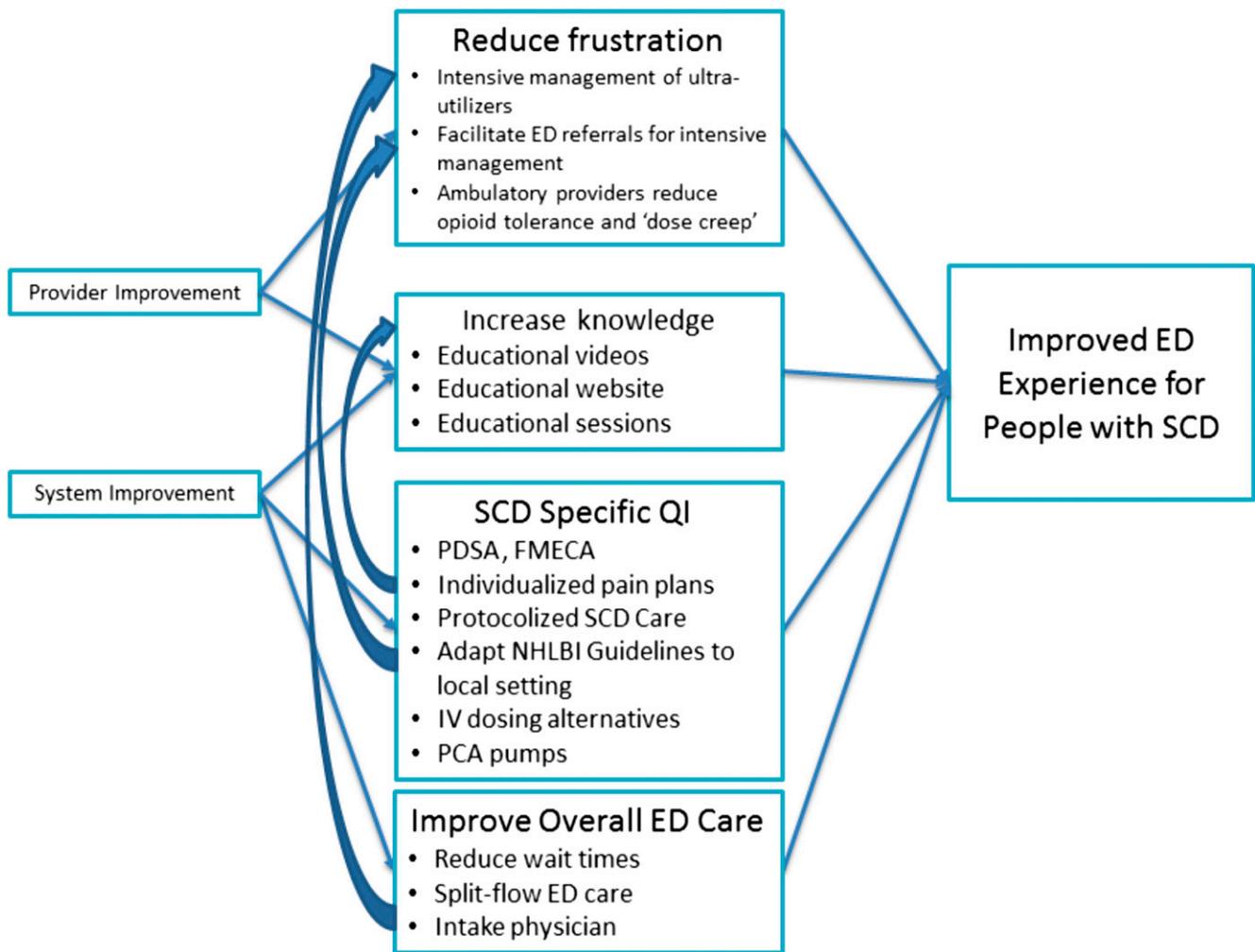


Figure 1. Elements of improving ED experience for people with SCD pain. SCD-specific quality improvement (QI) and general ED systems improvement can work to increase provider knowledge and decrease provider frustration, which ultimately lead to better patient-centered ED outcomes for acute SCD pain. FMECA, Failure Mode, Effects and Criticality Analysis; PDSA, plan-do-study-act.

provider frustration. Strategies to implement protocols and mitigate scarcity of resources are addressed in the latter 2 sections of this article.

In 2014, the National Heart, Lung, and Blood Institute (NHLBI) released guidelines for the care of SCD. Included among this document (freely available to the public)⁹ were recommendations for the management of acute sickle cell pain in the ED. The guidelines include several key elements. Patients with SCD should be assessed and triaged rapidly and assigned high priority for evaluation by a physician. The initial assessment should focus on determining if the patient is experiencing sickle cell pain and if there are other complicating medical issues that need to be addressed (eg, infections, cardiopulmonary emergencies). For patients experiencing only sickle cell pain, a weight-based or individualized (if available) pain management plan should be used, which involves rapid administration of an opioid analgesic followed by re-assessment and repeat dosing every 15 to 30 minutes “until the pain is controlled.” The NHLBI guidelines provide a framework to understand the elements of ideal emergency sickle cell pain care; however, they do not provide guidance on barriers and facilitators to achieving these ideals in the complex system of the ED. The following discussion covers 4 tenets of implementing guideline-adherent emergency sickle cell

care gleaned from the available literature and continuous quality improvement efforts at our institution (Figure 1).

Negative provider attitudes can be managed with a combination of education and intensive management of challenging patients

Negative provider attitudes toward individuals living with SCD are a barrier to the delivery of guideline-adherent pain care in the ED.¹⁰⁻¹² Negative attitudes among emergency providers are pervasive and include perceptions that people with SCD are addicted to opioids, that they exaggerate their pain, and that they are dishonest, uncooperative, manipulative, and frustrating to manage.¹³⁻¹⁵ For example, it has been shown that 2% to 5% of the SCD population (similar to the general population) meets criteria for a diagnosis of opioid addiction, but in 1 study, 46% of emergency physicians reported the belief that >10% of individuals with SCD are addicted.¹⁶ Emergency physicians’ negative attitudes likely affect the care they deliver. In the largest survey of emergency providers, individuals in the highest quartile of negative attitudes were significantly less likely to be willing to repeat doses of opioids for sickle cell pain.¹⁵

Negative provider attitudes also fracture the therapeutic relationship between health care provider and patient. Patients may perceive bias

on the part of providers, causing mistrust and stress, which is associated with more pain.^{17,18} Patients will then often take an adversarial position with health care providers, which can in turn increase provider frustration and bias.

One well-studied approach to improving negative provider attitudes is education. Educational sessions for health care staff have been shown to improve provider attitudes,¹⁹ but the most established approach involves the use of a short educational video. Haywood et al²⁰ used an 8-minute video in a pre–post intervention study of physicians and nurses who take care of people with SCD. The video features Dr. Sophie Lanzkron, who runs the adult SCD clinic at Johns Hopkins, and 3 adult patients with SCD who discuss their experiences while seeking treatment of acute exacerbations of sickle cell pain. There was a 9-point improvement in negative provider attitudes (using a validated instrument called the General Perceptions About Sickle Cell Patients Scale) after viewing the video. It is speculated that the video’s effectiveness is rooted in the fact that it allows emergency providers to see people with SCD outside of the ED when they are not in pain.^{20,21} The video also underscores the concept that most people with SCD do not want to come to the ED for treatment and that they do so only when other options have been exhausted.

Negative provider attitudes, particularly the perceptions that patients with SCD exaggerate their pain, are addicted to opioids, and are manipulative, can only partially be managed with education. Another indispensable component of managing negative emergency provider attitudes includes intensive management (a term initially used in SCD by Koch et al²² to describe a multilevel intervention for ultra-utilizers—variably defined as >4–12 ED visits/y) for the few patients who are addicted to opioids and have behavioral problems. It is well-known that a small subset of individuals with SCD account for the majority of ED visits and that these individuals are more likely to have substance abuse and behavioral disorders and be tolerant to opioids.²³ For many individuals who fit into this category of “ultra-utilizers,” intravenous (IV) opioids may not be in the patients’ best interest every time they present to the hospital with pain.

Although there may be a small subset of individuals with SCD for whom IV (and in some cases all) opioids should be withheld, we feel firmly at our center that an emergency provider should never have to make this judgment about a patient in the midst of a shift. EDs are chaotic, and providers make most of their decisions with partial or incomplete information. There are reasons other than addiction for why a patient may begin to use the ED more frequently for SCD pain (particularly bony etiologies such as undiagnosed avascular necrosis, bone infections, and infarcts). If an emergency provider mistakenly labels an individual with SCD as an ultra-utilizer (with all the associated negative connotations that this label carries), the results can have a substantial negative impact on that individual’s physical and emotional well-being.^{18,24}

Anecdotally, we have found that giving emergency providers a clear pathway (eg, a designated contact person to e-mail or call) to refer cases suspected of inappropriate ultra-utilization and showing providers that ultra-utilizers are being intensively managed by a sickle cell team, may help reduce emergency provider’s tendency to make judgments during ED shifts regarding which patients with SCD should and should not receive IV opioids. Intensive management strategies have been shown to reduce ED visits, admission rates, and hospital length of stay without an increase in adverse events⁵; it likely also reduces emergency provider frustration. Intensive management

typically involves performing a comprehensive biopsychosocial assessment to identify factors that are contributing to an individual’s high rates of utilization.^{5,22} Particular attention should be paid not to miss silent cerebral infarcts, which can limit an individual’s executive functioning and ability to follow treatment plans. Extensive efforts are made to meet the patient’s unmet needs and to initiate treatment of any addiction or behavioral issues. If the individual continually refuses to participate in a plan to reduce his or her ED utilization, only then are treatment plans put into place that involve withholding IV opioids in the ED.

Although the effect of intensive management of high utilizers on physician attitudes has not been quantified, it has been our experience that education and intensive management work well together to reduce negative emergency provider attitudes. At our institution, emergency providers are given contact information for the care management team that handles individuals when ultra-utilization is a concern. These providers are educated to trust patient’s reports of pain in the ED and never to withhold or reduce opioids for concerns about exaggeration or addiction. Instead, they are instructed to contact the care management team if they are concerned about a particular individual and are reassured that their concerns will be thoroughly investigated and addressed if necessary. Although this model has been successful at our institution and others, many institutions do not have a sickle cell care management team. However, in such institutions, other resources can be leveraged. Most hospitals have care management teams for high-risk patients, and these resources should be used whenever necessary for SCD.

Use strategies to improve time-to-first-dose: education, efficiency, and IV alternatives

ED patient satisfaction is strongly correlated with wait times, specifically “door-to-room” and “door-to-doctor” times.^{19,25,26} Taken within the context that the NLHBI guidelines recommend patients with acute SCD pain should receive their first analgesic within 60 minutes of arrival to the ED, strategies to reduce time-to-first-dose of pain medicine are highly beneficial.

Education, the bedrock of any ED quality improvement initiative for SCD, is an essential first step to reducing time-to-first-dose for SCD pain. Nurses, physicians, and other providers receive variable amounts of SCD education during their training and may have knowledge gaps. Educational modules should teach providers about SCD pathophysiology, clinical complications, and the nature and treatment of SCD pain. Emergency provider education should also focus specifically on the potential for individuals with SCD to develop life-threatening complications (acute chest syndrome, sepsis, pulmonary embolism, splenic sequestration, and stroke) and to clinically decompensate rapidly. Such educational modules can reduce time-to-first-dose by helping providers understand how severe sickle cell pain can be, and that prompt evaluation and treatment may also prevent other life-threatening complications. The website sickleemergency.duke.edu provides short educational modules tailored specifically to emergency providers.

Strategies to reduce ED wait times by improving overall ED efficiency can benefit SCD patients and can work to reduce and eliminate other health care disparities in the ED, as individuals with SCD are already known to experience wait times 25% longer than individuals with other conditions.^{21,27} Efficiency measures include split patient flow (where patients are assigned to different areas based

on acuity) and using an intake physician (where a physician is stationed in the triage area) to prescribe early analgesics. Although split flow models may improve overall ED efficiency, they are complicated by the NHLBI recommendation that all patients with SCD receive an emergency severity index classification of 2 or lower, as some emergency severity index–2 patients will wait longer to be seen in split flow models. Ultimately, split flow is likely beneficial to SCD care because it reduces the time from arrival to evaluation by a provider. Intake physician models can also benefit patients with SCD pain, as the physician can write orders for opioid analgesics immediately after the patient is triaged by a nurse.

Using alternatives to intravenous (IV) administration is a key strategy for reducing time-to-first dose, because patients sometimes wait hours in the ED before an IV can be inserted. Delays to IV placement occur because of surges in ED patient volume (ie, nurses are too busy) or because of difficulties in obtaining IV access that require the use of ultrasound guidance by a physician. In 1 study of children, intranasal fentanyl (as part of an overall SCD protocol) was associated with dramatic reductions in time to the first and second dose of analgesic.²⁸ First dose subcutaneous opioids (an approach recommended by the NHLBI guidelines) and first dose oral opioids are also excellent strategies to mitigate delays in IV placement.^{29–31} Intramuscular dosing should be avoided because it is more painful and provides no pharmacologic advantage over subcutaneous administration.

Beyond the first dose: individualized care plans, quality improvement, outpatient opioid reduction, and patient-controlled analgesia

The NHLBI guidelines recommend repeated assessments and doses of opioids every 15 to 30 minutes in the ED until sickle cell pain is relieved or reduced. Adherence to the NHLBI guidelines is associated with reductions in ED length of stay, fewer admissions to the hospital, and improved patient satisfaction,^{28,32} but adherence (in particular the recommendation to repeat analgesic doses every 30 minutes) is challenging. Quality improvement efforts show that reducing time-to-first-dose is more easily achieved than increasing the frequency of assessments and doses, particularly in adult settings.^{33,34} The best available literature and our experience suggests that 3 strategies are helpful in this area: rigorous quality improvement initiatives, use of individualized dosing plans, and aggressive outpatient efforts to reduce opioid use and limit opioid tolerance. Patient-controlled analgesia (PCA) devices may also be useful in a small subset of EDs with the resources available to use them properly.

Quality improvement is the only established approach to truly increase the frequency of assessments and analgesic doses in the ED. Several reports of quality improvement efforts in the ED exist, with most using a Failure Mode, Effects and Criticality Analysis in the context of a “plan-do-study-act” quality improvement program.^{28,34} Ultimately, quality improvement efforts should aim to establish and implement a protocol for the treatment of SCD pain, adapted from the NHLBI guidelines. Such efforts serve to reduce variability in care between providers and, in some situations, improve metrics of guideline adherence. Unfortunately, even published SCD quality improvement initiatives regularly fail; however, the following strategies may facilitate success.

Resource limitations in the ED may make it impossible to reassess and redose analgesics every 30 minutes; consequently, strategies that reduce the need for repeat doses may be the only effective approach. Individualized dosing plans (in which a SCD expert creates a dosing

plan for an individual based on his or her home opioid consumption and what doses have worked well during previous ED visits) typically result in higher opioid doses than would otherwise be given according to weight. Because these individualized dosing plans often result in more appropriate opioid dosing, they can reduce the need for repeat opioid doses. Most electronic medical record systems have the capability to store individualized dosing plans for rapid retrieval by the treating emergency provider.

A second strategy involves aggressive efforts by outpatient SCD physicians to reduce long-term opioid use and tolerance (discussed elsewhere in this issue). During an acute SCD pain episode, opioids are almost always medically indicated. As the pain episode resolves, aggressive outpatient efforts to titrate down opioids can limit the potential for increasing opioid tolerance. By limiting opioid tolerance in this manner, patients may achieve pain relief in the acute setting with fewer doses of opioids, thus mitigating the challenges of repeat dosing with short, rapid intervals.

PCA is a promising strategy to safely provide repeat doses of analgesics without straining available ED resources. Several observational studies have reported that PCAs can be used in the ED^{35,36} and can minimize gaps in pain management as patients are transferred from ED to inpatient status. However, in our experience, we have found that if PCA dose regimens are not carefully written, use of PCAs may actually result in worsened pain management. Because many patients with SCD are opioid tolerant, they may require high-dose PCA settings that many emergency providers are not comfortable ordering. Furthermore, PCAs deliver small frequent doses that may take longer to attain high plasma levels and achieve pain relief. This scenario may be mitigated by using individualized PCA plans or by working in consultation with a board-certified pain management physician to ensure proper PCA settings.

Patient safety and clinical pearls for ED providers

Very few data are available regarding patient safety for ED management of SCD pain. Although patients tend to focus on problems with stigmatization and inadequate analgesia, providers may focus on safety and avoiding harm. One retrospective study showed that although high-dose opioids can be used safely in the ED, patients are more likely to experience abnormal or dangerous vital sign abnormalities with increasing opioid doses.³⁷ In addition to the strategies discussed earlier, the following clinical pearls help ED providers provide safe, high-quality pain care to ED patients with SCD.

- **SCD pain is a diagnosis of exclusion**—Emergency providers are trained to form ranked differential diagnoses and to immediately rule-out potentially life-threatening causes for a patient’s symptoms. Providers often take cognitive shortcuts for patients with SCD pain, assuming that their symptoms are simply a manifestation of their disease without carefully ruling out other dangerous causes of pain. ED providers should be reminded to consider all the possible causes of a patient’s symptoms (eg, for lower abdominal pain, consider appendicitis and ovarian torsion; for chest pain, consider pulmonary embolism) and to avoid premature diagnostic closure.
- **Pulse oximetry and cardiac monitoring during the initial analgesic phase**—Although the safety of high-dose opioids for SCD pain has been shown,³⁷ there is still a small risk of serious adverse events (eg, respiratory depression, hypotension, bradycardia), especially if there is an unrecognized secondary etiology for the patient’s pain (eg, infection, pulmonary embolism). In addition, many patients with SCD are taking multiple QT-interval–prolonging

medications, which can increase the risk of arrhythmias. Telemetry and continuous pulse oximetry monitoring in the ED may facilitate more rapid recognition of rare adverse events and reduce the potential for patient injury.

- **Hydration does not cure crisis; use gentle hypotonic fluids unless otherwise indicated**—Many emergency providers are taught to bolus isotonic crystalloid as a treatment for acute SCD pain. There is indirect evidence that this approach may increase the likelihood of developing acute chest syndrome.³⁸ Isotonic fluids should be used when indicated (eg, hypovolemia, vasodilation), and gentle administration of hypotonic fluids should be used otherwise.
- **Creatinine does not accurately reflect glomerular filtration rate in SCD**—Many emergency providers are unaware that patients with SCD develop hyposthenuria from renal microinfarcts, which lowers serum creatinine levels but does not reflect an increase in glomerular filtration rate. Use of nephrotoxic and renally cleared medications (eg, nonsteroidal anti-inflammatory drugs, meperidine) should be minimized, and use of IV contrast for CT scanning should also be minimized if possible.
- **Exchange or simple transfusion should be considered for all forms of critical illness in SCD, including but not limited to acute chest syndrome**—Emergency providers are often confused about the management of acute chest syndrome because there is significant diagnostic overlap with pneumonia. Ultimately, emergency providers need to know that all patients with SCD who are critically ill, or continuing to deteriorate despite supportive therapy, should be considered for exchange transfusion and that it is of little consequence whether the patient has acute chest syndrome or a different, life-threatening event. Emergency providers should be encouraged to involve hematology consultants early to consider exchange or simple transfusion when a patient does not respond to initial supportive care.³⁸

Conclusions

Management of SCD pain in the ED is challenging, and research is needed at several levels to improve care delivery. Systems-level research can identify better strategies to improve ED care using existing resources. There are also emerging therapies, including the use of novel agents (eg, selectin inhibitors) as well as existing agents (ketamine), that may provide much-needed alternatives to opioids. Using the strategies outlined here can maximize analgesia and safely improve patient satisfaction.

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