Pain Management for Sickle Cell Disease in the Pediatric Emergency Department: Medications and Hospitalization Trends

Chantel Cacciotti, MD1,2, Sarah Vaiselbuh, MD1, and Eleny Romanos-Sirakis, MD1

Abstract
The majority of emergency department (ED) visits and hospitalizations for patients with sickle cell disease (SCD) are pain related. Adequate and timely pain management may improve quality of life and prevent worsening morbidities. We conducted a retrospective chart review of pediatric patients with SCD seen in the ED, selected by sickle cell–related ICD-9 codes. A total of 176 encounters were reviewed from 47 patients to record ED pain management and hospitalization trends. Mean time to pain medication administration was 63 minutes. Patients received combination (nonsteroidal anti-inflammatory drug [NSAID] + narcotic) pain medications for initial treatment at a minority of ED encounters (19%). A higher percentage of patients who received narcotics alone as initial treatment were hospitalized as compared with those who received combination treatment initially (P= 0.0085). Improved patient education regarding home pain management as well as standardized ED guidelines for assessment and treatment of sickle cell pain may result in superior and more consistent patient care.

Keywords
sickle cell disease, vaso-occlusive event, home pain management

Introduction
Sickle cell disease (SCD) is an inherited multisystem disease affecting approximately 72,000 to 98,000 children and adults in the United States, accounting for early mortality.1 The majority of emergency department (ED) visits and hospitalizations for SCD are related to painful vaso-occlusive events (VOEs).2 Sickle cell pain is unpredictable and triggers may not be evident. Known risk factors for sickle cell acute pain include physiological, psychological, and/or physical stressors.3

Pain in SCD is a heterogeneous and complex entity. Inadequate pain management in sickle cell patients is associated with increased morbidity, with potential complications in nearly all body systems, and decreases quality of life in patients of all ages.4 Tissue damage caused by vaso-occlusion triggers a complex cascade of events leading to acute pain, which may become chronic. In addition, the vaso-occlusion leads to vascular inflammation.5 Adequate and timely pain management improves quality of life, and rapid evaluation of pain is critical to ensure quick pain relief and prevention of complications.

Despite the severity of SCD-related VOE and acute pain episodes, pediatric ED analgesic guidelines for SCD pain are not universal. Our goal was to retrospectively review the documentation of pain scales and the pain regimens used for patients with SCD-related pain in the ED at our institution. In addition, we aimed to determine factors that influence the type of pain treatment regimen prescribed and the amount of time to treatment. We also evaluated the relationship between pain management and rates of hospitalization and length of stay.

Materials and Methods

Study Design and Setting
A single-institution, retrospective review of medical records was used to extract data, including the following variables: demographics (gender, age), diagnosis, time from triage to initial analgesic medication, analgesic

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agents, time to reassessment after initial pain medication was administered, pain scale at initial presentation and reassessment, ED physician caring for the patient (pediatric or adult trained), disposition (admission to the hospital or discharge from the ED), and length of stay if hospitalized.

**Selection of Participants**

Our study cohort was selected using the following data collection points: pediatric patients with SCD, 0 to 21 years old, presenting to the ED with an acute pain-related complaint, determined by ICD-9–related SCD codes—sickle cell with crisis (282.60, 282.62, 282.64, 282.69) and acute chest (517.3)—between October 2004 and September 2013 were included. A total of 176 ED visits were reviewed from 47 unique patients (range of 1-18 ED encounters/patient).

**Methods and Measurements**

All pain scores were extracted as documented in the electronic medical record using a pain scale from 0 to 10. Pain scales were subcategorized into mild (pain scale 1-3), moderate (pain scale 4-6), or severe (pain scale 7-10). The scales were extrapolated for some patients based on qualitative description of pain. Time to administration of pain medication was calculated starting from ED triage. Analgesic agents were categorized as narcotic (Morphine, Dilaudid, Percocet, or Oxycodone), nonsteroidal anti-inflammatory drug (NSAID; Ibuprofen, Ketorolac), or combination (NSAID and narcotic).

Pain scales utilized included Wong-Baker FACES pain scale readings or Numerical Rating Scale.5,6 The numerical rating scale involves the patient stating his/her level of pain on a scale of 1 to 10, with 0 representing no pain and 10 the worst pain. The Wong-Baker FACES pain scale utilizes faces to express pain while asking the patient to identify the face that best describes his/her pain.5,6

**Analysis**

Descriptive statistics such as the mean, SD, and median were calculated on the continuous variables. Frequencies and percentages were tabulated from the categorical and ordinal variables. The Mann-Whitney or Kruskal-Wallis test, as appropriate, was used to compare groups on a number of selected continuous variables. Significant omnibus results were explored further with pairwise comparisons, and a Bonferroni adjustment was made for multiple testing. The association between categorical and ordinal variables such as pain scale, discharge status, medications, and so on was explored using the $\chi^2$ or Fisher’s exact test, as appropriate. The Spearman correlation was computed to test the relationship between pairs of variables related to several aspects of pain. An $\alpha$ level of < .05 was used to declare significance. Statistical analysis was performed using SAS version 9.3 (SAS Institute Inc, Cary, NC).

**Results**

**Demographics**

The age, diagnosis, and gender of our patients are depicted in Table 1. The mean age of patients was 12.5 years (range of 6 months to 21 years). The majority of patients had a documented diagnosis of hemoglobin SS.

**Pain Assessment**

A total of 167/176 (94%) encounters had pain scales documented at triage; 28/167 (17%) patients reported mild pain (pain scale 1-3), 51/167 (30%) moderate pain (4-6), and 88/167 (53%) severe pain (7-10). After initial analgesic intervention, 8/166 (5%) reported no pain, 51/166 (31%) mild pain, 47/166 (8%) moderate pain, and 31/166 (19%) severe pain (Table 2). On reassessment (after initial analgesic medication intervention), the percentage of patients reporting severe pain (pain scale 7-10) decreased from 53% to 19%.

**Time to Pain Medication**

The mean time (±SD) from triage to pain medication administration was 63 (±46) minutes (range = 10-300 minutes), and the mean time to reassessment after pain medication was 100 (±63) minutes. The length of time to initial pain medication administration was not significantly associated with the intensity of the initial pain scale, patient age, or type of provider (adult vs pediatric-trained ED staff).

**Type of Analgesic Administration**

At 19% (32/168) of encounters, patients received combination treatment as initial pain management (both

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**Table 1. Demographics.**

<table>
<thead>
<tr>
<th>Gender</th>
<th>Percentage male (n)</th>
<th>46% (78)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Percentage female (n)</td>
<td>54% (91)</td>
</tr>
<tr>
<td>Mean (years)</td>
<td></td>
<td>12.5</td>
</tr>
<tr>
<td>Sickle cell subtype</td>
<td>Percentage Hgb SS, sickle cell SS disease (n)</td>
<td>93% (148)</td>
</tr>
<tr>
<td>Percentage Hgb SC, sickle cell SC disease (n)</td>
<td>7% (11)</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviation: Hgb, hemoglobin.
NSAID and narcotic); 48% (81/168) received NSAID alone, and 33% (55/168) received narcotics alone as initial treatment (Table 3). Patients received combination treatment over the course of the entire ED visit at 24% (40/168) of encounters.

A significant association was noted between initial pain scale and initial pain medication given in the ED ($P < .0001$; Figure 1). Not surprisingly, patients who received initial treatment with NSAIDs alone had a lower median initial numerical pain scale compared with the other 2 treatment groups. However, after running a pairwise comparison, no significant association between the severity of the pain scale and initial treatment with narcotic alone versus combination treatment was found.

Patient age was also significantly associated with the initial medication given ($P < .0001$; Table 3). Patients who received narcotics alone as initial pain medication were older (16.6 years old) as compared with the NSAID-only and combination treatment groups (9.6 and 13.4 years old, respectively).

The type of training of the ED provider (pediatric vs adult training) was significantly associated with the type of initial pain medication provided ($P = .041$). The pediatric-trained ED providers more commonly administered NSAIDs as the initial treatment (64.4% of encounters) as compared with narcotics (20.6%) or combination treatment (15%). Adult-trained ED providers administered narcotics as initial pain medications at 39% of encounters, NSAIDs alone at 42.9% of encounters, and combination treatment at 18.1% of encounters. Among patients who received narcotics as initial pain treatment, 65.2% were treated by adult-trained providers versus 32.6% who were treated by pediatric-trained providers.

### Hospitalization

The type of initial pain medication administered was significantly associated ($P < .0001$) with hospitalization; higher admission rates were noted in patients who received narcotics as initial pain medication in comparison to either NSAIDs or combination treatments (Figure 2). Among admitted patients, 27% received NSAIDs alone, 56% received narcotics alone, and 17% received combination treatment (NSAID + narcotic) as initial treatment in the ED. Patients who received narcotics alone were more likely to be admitted in comparison to those who received combination treatment initially ($P = .0085$). Also, 64% of patients who received narcotics alone as initial treatment were hospitalized from the ED as compared with 33% of patients who received combination treatment initially. Among patients who received initial combination pain treatment of NSAIDs and narcotics, the majority (66.7%) were discharged home from the ED.

Not surprisingly, the highest proportion of admitted patients reported severe pain at triage. The same trend confirmed itself at reassessment, with a higher proportion of patients with severe pain remaining at reassessment being hospitalized ($P < .0001$; Figure 3). Of note, among patients with severe pain at reassessment, 35% were discharged.

### Length of Hospitalization

No significant association was noted between duration of hospitalization and the following variables: initial pain scale, time to initial pain medication, or type of initial pain medication. Among admitted patients, the median duration of hospitalization was 3 days (range = 1-11 days).

### Limitations

We performed a retrospective chart review, which is subject to the limitations of provided documentation. Documented pain scales or identification of pain as mild, moderate, or severe was subjective, and pain scales were extrapolated for some patients based on qualitative description of pain. Numerical pain scales were used intermingled with Wong-Baker Face pain scales regardless of patient’s age, at the discretion of the ED physician. The choice of first-line analgesics as well as the timing and use of combination analgesics (initial or at reassessment) were also at the sole discretion of the ED physician.
Discussion
Varied guidelines have been published to optimize pain management for patients with sickle cell pain.\textsuperscript{7,8} Recommendations indicate that analgesic medications should be provided within 30 minutes of arrival to the hospital. Time to pain medication is often delayed. Emergency room crowding and radiology testing are among the factors that contribute to delays in pain medication administration.\textsuperscript{9,10} In addition, only 20% of adult ED physicians and 9% of pediatric ED physicians report utilizing protocols when managing sickle cell–related pain, and studies have shown considerable variability in the treatment of sickle cell pain in the ED.\textsuperscript{11}
The etiology of SCD pain is multifactorial; SCD pain encompasses both inflammation and infarction/tissue damage. Optimal pharmacological intervention should aim at targeting both the inflammatory and tissue damage components of the pain to optimize treatment. A combination regimen of NSAIDs (anti-inflammatory) and narcotics (pain of tissue necrosis) addresses this pathophysiology. Interestingly, in our study, whereas patients with mild pain most commonly received NSAIDs, among patients with moderate/severe pain, we found no significant association between pain scale and initial pain treatment with narcotics versus combination medications. In addition, patients who received combination treatment had lower admission rates as compared with patients initially treated with narcotics alone. It is understandable that the patients treated with narcotics would have higher admission rates as compared with those treated with NSAIDS alone, given the association between lower pain scale and treatment with NSAIDs alone. However, the higher admission rates among those treated with narcotics alone as compared with combination treatment of narcotics and NSAIDS requires further investigation; we surmise that because the combination treatment targets 2 mechanisms of pain in SCD, combination treatment in the ED may play a role in decreasing the need for admission in some cases. Aggressive titration of the pain regimen until relief is achieved in a day hospital setting has been shown to decrease admissions in patients with SCD presenting with pain.\textsuperscript{4} Combination medication may contribute to optimizing pain management in the ED and, thus, may decrease admission rates in a fashion similar to that of the day hospital model.

Several pharmacological options exist for the treatment of acute pain episodes; analgesic options include NSAIDs, acetaminophen, and opioids. Acetaminophen and NSAIDs are generally used as first-line management of less severe pain, whereas oral or intravenous (IV) opioids can be added for moderate to severe pain.\textsuperscript{12} Disparities exist in the choice of pain management regimens of sickle cell VOE in the ED, possibly because of treatment by a diversity of ED staff with different approaches to pain control.\textsuperscript{11}

Initial pain scale assessment in the ED should aim at providing rapid pain control in sickle cell patients. Our single institution study demonstrated a median time to pain medication of 60 minutes and median time of 90 minutes to reassessment after pain medication was given. We found that the time to initial pain medication administration was not associated with the intensity of the pain scale, suggesting that higher pain scales did not elicit a more rapid response of ED staff to give treatment.

Education and appropriate patient-specific home pain management education are required for optimal management of this complex disease.\textsuperscript{2} In this study, 17% of patients were seen in the ED with reports of mild pain (pain scale = 1-3). Patients with lower pain scales were
more likely to be treated with NSAIDs alone. This popu-
lation presenting with mild pain would benefit from fur-
ther education regarding pain management at home and
individualized pain plans to prevent some potentially
unnecessary visits to the ED. In addition, outpatient/ 
home management of SCD pain is beneficial because it
provides patient autonomy as well as results in more fre-
cent interaction with the primary hematologist who,
then, can guide and help in decision making regarding
ED visits. It is prudent for patients to be educated to
know when a visit to the ED is beneficial and necessary
to avoid progressive morbidity.10

Conclusion

Time to initial treatment and reassessment of pain var-
ded and was often prolonged. Providers were more
likely to treat lower pain scales with NSAIDs alone;
these patients may benefit from education regarding
home pain regimens that may prevent potentially
unnecessary ED visits. Initial treatments varied; the
decision to provide combination treatment to only a
small proportion of patients warrants further investiga-
tion. Patients who received combination treatment were
less likely to be admitted from the ED for further pain
management. Approximately one-third of patients with
severe pain at reassessment were discharged, which
also warrants further evaluation. Comprehensive ED
guidelines for assessment and treatment of SCD pain
may contribute to optimal and consistent care. Improved
pain management may help prevent hospitalizations
and morbidity associated with sickle cell pain.

Future Directions

With the goal of appropriate assessment and increased
outpatient management of mild SCD pain, individual-
ized outpatient pain management plans (“Stop Pain”
Program) were created to further educate and encourage
patients and their families to become more actively
involved in their own care. With this initiative, we hope
to reduce unnecessary ED visits, improve home pain
management, and increase patient autonomy. In addi-
tion, further collaboration between all providers who
treat patients with SCD-related pain and the further
development of guidelines for pain management in the
ED may lead to improved quality of life and decreased
morbidity for this patient population.

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Romano-Sirakis, MD, each of whom have truly contributed in
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The original manuscript was written by Dr. Cacciotti. All authors
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