

Children's Mercy Kansas City

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Clinical Pathways

Evidence-Based Practice Collaborative

10-2023

Sickle Cell Disease: Management of Acute Pain

Children's Mercy Kansas City

These guidelines do not establish a standard of care to be followed in every case. It is recognized that each case is different and those individuals involved in providing health care are expected to use their judgment in determining what is in the best interests of the patient based on the circumstances existing at the time. It is impossible to anticipate all possible situations that may exist and to prepare guidelines for each. Accordingly, these guidelines should guide care with the understanding that departures from them may be required at times.

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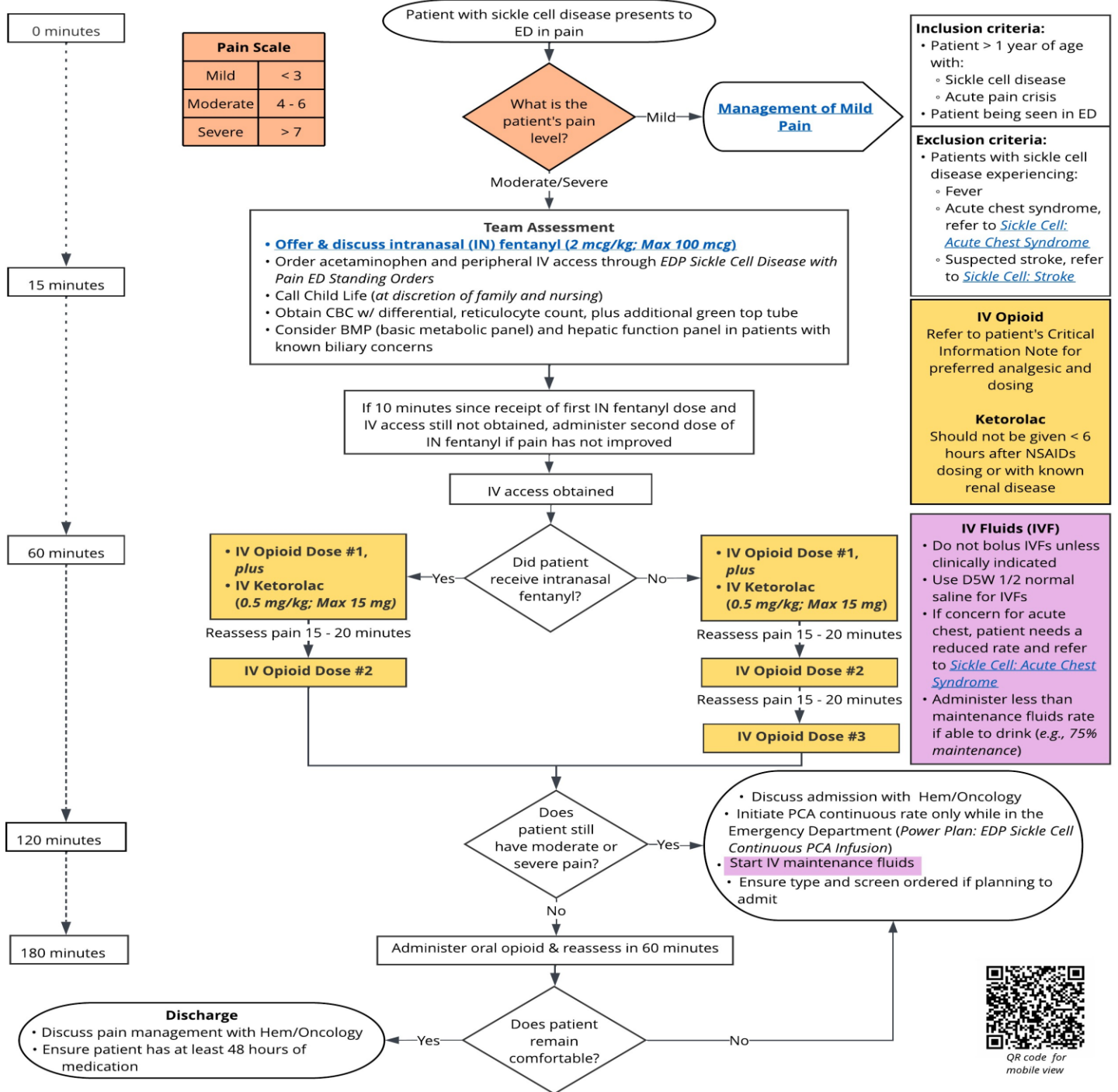


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Sickle Cell Disease: Management of Acute Pain Clinical Pathway Synopsis

Sickle Cell Disease: Management of Acute Pain



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Sickle Cell Disease: Management of Mild Pain

- Inclusion criteria:**
- Patient >1 year of age with:
 - Sickle cell disease
 - Acute pain crisis
 - Patient being seen in ED
- Exclusion criteria:**
- Patients with sickle cell disease experiencing:
 - Fever
 - Acute chest syndrome, refer to [Sickle Cell: Acute Chest Syndrome](#)
 - Suspected stroke, refer to [Sickle Cell: Stroke](#)

Patient with sickle cell disease presents to ED in need of management for mild pain

Administer ibuprofen and home oral opioid as listed in Critical Information Note

Reassess pain 60 minutes

Has the patient's pain improved and/or is family comfortable with discharge home?

Follow **Moderate/Severe** Pain

Discharge

- Discuss pain management with Hem/Oncology
- Advise patient/family of the time the next doses are due
- Administer acetaminophen if > 6 hours since last dose
- Continue home oral analgesic regimen
- Ensure patient has at least 48 hours of medication

Pain Scale	
Mild	< 3
Moderate	4 - 6
Severe	> 7



QR code for mobile view

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Objective of Clinical Pathway

To provide care standards for patients diagnosed with sickle cell disease, who present to the emergency department for management of an acute pain episode. The Sickle Cell Disease: Management of Acute Pain Clinical Pathway provides guidance regarding recommended assessment and treatment to minimize variation of care and decrease the time to administration of first analgesic.

Background

Children with sickle cell disease experience acute pain episodes often requiring emergency department visits and for which optimal pain management requires the coordinated efforts of an interprofessional team (Brandow et al., 2020; Brousseau et al., 2020; Puri et al., 2018). Guidelines recommend that when treating children with sickle cell disease who are experiencing an acute pain episode the first dose of pain medication be administered within 30 minutes of triage in the emergency department or 60 minutes following emergency department arrival (Brandow et al., 2020; National Heart, Lung, and Blood Institute, 2014). Historically, time to first analgesic administration varies where children in an acute pain crisis do not receive their first dose of pain medication within the recommended time (Brousseau et al., 2020; Lin et al., 2016). Delays are noted regardless of whether an institution has an established sickle cell acute pain protocol in place to expedite triage and initiate intravenous (IV) access (Brousseau et al., 2020; Fein et al., 2017).

Due to delays in time to first analgesic, particularly for children experiencing sickle cell pain episodes that are moderate to severe, providers have sought alternative methods for first analgesic delivery to address this care gap (Brousseau et al., 2020; Fein et al., 2017; Kavanagh et al., 2015; Kelly et al., 2018; Puri et al., 2018; Rees et al., 2023). Intranasal fentanyl is a treatment that has demonstrated benefits for use in the pediatric emergency department setting to close the care gap for children experiencing moderate to severe sickle cell pain episodes (Brousseau et al., 2020; Fein et al., 2017; Kavanagh et al., 2015; Kelly et al., 2018; Puri et al., 2018; Rees et al., 2023). The Sickle Cell Disease: Management of Acute Pain Clinical Pathway aims to address the time to first analgesic in the emergency department for children experiencing an acute sickle cell pain episode through the development of an evidence-based clinical pathway focusing on assessment, treatment, and reassessment through an interprofessional team approach.

Target Users

- Physicians (Emergency Department, Fellows, Resident Physicians)
- Nurse Practitioners
- Nurses
- Pharmacy

Target Population

Inclusion Criteria

- Patients > 1 year of age with:
 - Sickle cell disease
 - Acute pain crisis
- Patient being seen in the emergency department (ED)

Exclusion Criteria

- Patients with sickle cell disease experiencing:
 - Fever
 - Acute chest syndrome, refer to [Sickle Cell: Acute Chest Syndrome](#)
 - Suspected stroke, refer to [Sickle Cell: Stroke](#)

AGREE II

The American Society of Hematology 2020 Guidelines for Sickle Cell Disease: Management of Acute and Chronic Pain provided guidance to the Sickle Cell Disease: Management of Acute Pain committee (Brandow et al., 2020). See Table 1 for AGREE II.

Table 1

AGREE II^a Summary for the American Society of Hematology 2020 Guidelines for Sickle Cell Disease: Management of Acute and Chronic Pain (Brandow et al., 2020)

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Domain	Percent Agreement	Percent Justification [^]
Scope and purpose	100%	The aim of the guideline, the clinical questions posed, and target populations were identified.
Stakeholder involvement	100%	The guideline was developed by the appropriate stakeholders and represents the views of its intended users.
Rigor of development	98%	The process used to gather and synthesize the evidence, the methods to formulate the recommendations and to update the guidelines were explicitly stated.
Clarity and presentation	100%	The guideline recommendations are clear, unambiguous, and easily identified; in addition, different management options are presented.
Applicability	97%	Barriers and facilitators to implementation, strategies to improve utilization and resource implications were addressed in the guideline.
Editorial independence	100%	The recommendations were not biased with competing interests.
Overall guideline assessment	99%	
See Practice Recommendations		

Note: Four EBP Scholars completed the AGREE II on this guideline.

[^]Percentage justification is an interpretation based on the Children’s Mercy EBP Department standards.

Practice Recommendations

Please refer to the American Society of Hematology 2020 Guidelines for Sickle Cell Disease: Management of Acute and Chronic Pain for assessment and treatment recommendations (Brandow et al., 2020).

Additional Questions Posed by the Clinical Pathway Committee

No clinical questions were posed for this review.

Updates from Previous Versions of the Clinical Pathway

- The Sickle Cell Disease: Management of Acute Pain Clinical Pathway is a newly developed evidence-based pathway of which there is not a previous version for comparison.

Recommendation Specific for Children’s Mercy

Children’s Mercy adopted the majority of the practice recommendations made by the American Society of Hematology 2020 Guidelines for Sickle Cell Disease; Management of Acute and Chronic Pain (Brandow et al., 2020). Variations/Additions include:

- Guidance provided by the American Society of Hematology (2020) recommends pain reassessment every 30-60 minutes for children with sickle cell disease presenting with acute pain in an acute care setting, whereas the Sickle Cell Disease: Management of Acute Pain Clinical Pathway Committee recommends reassessment for children experiencing moderate to severe pain every 15 – 20 minutes.

Measures

- Time to first analgesic
- Time to preferred opioid administration
- Time from intranasal fentanyl administration to IV opioid administration
- Time to disposition (discharge vs. admission)
- Percentage of patients requiring Vascular Access (VAT) consultation

Value Implications

The following improvements may increase value by reducing healthcare costs and non-monetary costs (e.g., missed school/work, loss of wages, stress) for patients and families and reducing costs and resource utilization for healthcare facilities.

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- Decreased time to first analgesic for children presenting to the ED with an acute exacerbation of sickle cell pain
- Decreased frequency of admission
- Decreased unwarranted variation in care

Organizational Barriers and Facilitators

Potential Barriers

- Variability of acceptable level of risk among physicians and nurses
- Increased concern about opioid misuse

Potential Facilitators

- Collaborative engagement across care continuum settings during clinical pathway development
- Standardized order set for the Emergency Department

Diversity/Equity/Inclusion

Our aim is to provide equitable care. These issues were discussed with the Committee, reviewed in the literature, and discussed prior to making any practice recommendations.

Power Plans

- EDP Sickle Cell with Pain ED Standing Orders
- EDP Sickle Cell Pain Crisis with or without Fever

Associated Policies

- Sickle Cell Disease with Pain Standing Order (updating; in process of approval)

Education Materials

- [Intranasal Fentanyl](#)
 - Intended to guide talking points for the provider and be a resource for families
 - Found as an Additional Tool associated with the clinical pathway
 - Available in English, Spanish, Swahili, Somali, and French

Clinical Pathway Preparation

This pathway was prepared by the Evidence Based Practice (EBP) Department in collaboration with the Sickle Cell Disease: Management of Acute Pain Clinical Pathway Committee composed of content experts at Children's Mercy Kansas City. The development of this pathway supports the Quality Excellence and Safety Division's initiative to promote care standardization that is evidenced by measured outcomes. If a conflict of interest is identified, the conflict will be disclosed next to the committee member's name.

Sickle Cell Disease: Management of Acute Pan Clinical Pathway Committee Members and Representation

- Allison Adam, MD | Emergency Department, Pediatric Emergency Medicine Fellow | Committee Chair
- Leslie Hueschen, MD | Emergency Department | Committee Chair/Member
- Michelle DePhillips, MD | Emergency Department | Committee Member
- Lina Patel, MD | Emergency Department | Committee Member
- Laurence Noisette, MD | Hematology/Oncology/BMT | Committee Member
- Shabnam Arsiwala, MD, FAAP | Hematology/Oncology/BMT | Committee Member
- Amy Johnson, MD, MBA | Hematology/Oncology, BMT, Pediatric Fellow | Committee Member
- Shailly Gaur, MD | Hematology/Oncology/BMT, Pediatric Fellow | Committee Member
- Charleen Cunningham, RN, MSN, CPN | Emergency Department, Assistant Director of Nursing | Committee Member
- Rae Kingsley, DNP, APRN, CPNP-AC/PC | Rheumatology/Pain Clinic/Rehabilitation for Amplified Pain Syndromes (RAPS) | Committee Member
- Ibad Siddiqi, PharmD | Pharmacy, Emergency Department Team Leader | Committee Member
- Chris Kennedy, MD | Emergency Department | Committee Member
- Sue Stamm, MSN, RN, CPNP, CPON | Clinical Practice and Quality | Committee Member

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Patient/Family Committee Member

- Jewel Akpan | Committee Member

EBP Committee Members

- Todd Glenski, MD, MSHA, FASA | Anesthesiology, Evidence Based Practice
- Kelli Ott, OTD, OTR/L | Evidence Based Practice

Clinical Pathway Development Funding

The development of this clinical pathway was underwritten by the following departments/divisions: Emergency Department, Hematology/Oncology, Rheumatology, Pharmacy, Clinical Practice and Quality, and Evidence Based Practice.

Conflict of Interest

The contributors to the Sickle Cell Disease: Management of Acute Pain Clinical Pathway have no conflicts of interest to disclose related to the subject matter or materials discussed.

Approval Process

- This clinical pathway was reviewed and approved by the Sickle Cell Disease: Management of Acute Pain Committee, content experts from related departments/divisions, and the EBP Department
- Pathways are reviewed and updated as necessary every 3 years within the EBP Department at CMKC. Content expert teams are involved with every review and update.

Review Requested

Department/Unit	Date Obtained
Emergency Department	October 2023
Hematology/Oncology/BMT	October 2023
Pain Clinic	October 2023
Pharmacy	October 2023
Clinical Practice and Quality	October 2023
Evidence Based Practice	September 2023

Version History

Date	Comments
October 2023	Version one (ED algorithm and Sickle Cell Disease: Management of Acute Pain synopsis developed, EDP power plans updated to align with clinical pathway, IN Fentanyl handout created)

Date for Next Review

- October 2026

Implementation & Follow-Up

- Once approved, the pathway was presented to appropriate care teams and implemented. Care measurements will be assessed and shared with appropriate care teams to determine if changes need to occur.
- Education tools were reviewed by health literacy and family advisory board member.
- Order sets/power plans consistent with recommendations were updated for the Emergency Department
- The policy was updated. This details a process for nursing staff for patients with sickle cell disease presenting in pain crisis to improve comfort and expedite care. The policy is in the process of approval by the Nursing Practice Council, Pharmacy & Therapeutics Committee, and Medical Executive Committee.
- Education was provided to all stakeholders:
 - Nursing units where the Sickle Cell Disease: Management of Acute Pain Clinical Pathway is used
 - Providers from Emergency Department and Hematology/Oncology/BMT
 - Resident physicians
- Additional institution-wide announcements were made via email, the hospital website, and relevant huddles.

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- Metrics will be assessed and shared with appropriate care teams to determine if changes need to occur.

Disclaimer

When evidence is lacking or inconclusive, options in care are provided in the supporting documents and the power plan(s) that accompany the clinical pathway.

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