Sickle Cell with Pain Crisis Guideline

Definition:

Children with sickle cell disease **(all genotypes)** presenting with acute pain event.

Relevance:

Vaso-occlusive pain crisis is the most common complication of sickle cell disease.[12] Pain events often lead to seeking medical care in the Emergency Department. Historically, there has been high variability in acute pain management within and between institutions for sickle cell disease, leading to delays in both time-to-first pain intervention and pain re-assessment, and resulting in poorer health outcomes.[12] The introduction of sickle cell pain protocols in the ED setting, both adult and pediatrics, has been shown to decrease time interval to first analgesic, improve appropriate weight based analgesic dosing, and decrease hospital admission rates.[12,13] As a result, there is ample literature to support an evidence based protocol approach to managing sickle cell pain in the acute care setting.[12, 13, 14] In addition, The 2020 American Society of Hematology guideline for management of acute and chronic pain recommends that individualized pain plans, with tailored medications and doses specific to a patient, may augment standard of care for pain management for patients with SCD.[14]

Incidence:

Sickle cell pain crisis is very common in both pediatric and adult patients; it is the most common reason for patients to seek medical attention. In one 2010 study, there were approximately 200,000 emergency department visits by children and adults with sickle cell disease, with 67 percent for pain alone. In comparison, visits for chest symptoms (pain, shortness of breath, cough) and fever accounted for only 20 and 6 percent of visits, respectively. Concurrent symptoms commonly occur; acute chest syndrome, stroke, and multi-organ failure can develop or occur simultaneously with an acute pain crisis (Darbari, Sheehan, Ballas 2020). In one adult study [Pain in Sickle Cell Epidemiology Study (PiSCES)], patients reported pain on 54.5% of the 31,017 days surveyed. Almost 30% of respondents had pain on more than 95% of the days surveyed. In the pediatric population, Dampier et al. studied children and adolescents (ages 6–21 years) with sickle cell disease for 18,377 days. Children commonly reported pain, with 514 distinct pain episodes occurring over 2592 days and 2326 nights. Acute pain is a known hallmark of sickle cell disease, with chronic pain often occurring frequently as well.[1-4]

Etiology:

Sickle cell disease is due to a single amino acid substitution in the gene encoding the β-globin subunit. Polymerization of deoxygenated sickle hemoglobin leads to decreased deformability of red blood cells. Through adhesive events among blood cells, these erythrocytes can obstruct the vasculature, producing pain, hemolytic anemia, organ injury, and early mortality. Although the molecular basis of SCD is well characterized, the complex mechanisms underlying vaso-occlusion have not been fully established. Preferential

adhesion of low-density SS-RBCs and reticulocytes in immediate postcapillary venules leads to trapping of the older, more dense, and misshapen SS-RBCs. Precapillary obstruction by a small number of dense SS-RBCs also contributes to VOC. Recent data indicates other blood cell elements that are not directly affected by the sickle cell mutation play a direct role in VOC. Theories have been proposed in which the process is viewed as multistep and multicellular cascade driven by inflammatory stimuli and the adherence of leukocytes.[5]

Differential Diagnosis:

Vaso-occlusive Crisis (VOC), Pneumonia, Pulmonary Embolism, Acute Chest Syndrome, Reactive Airway Disease, Asthma, Cardiomyopathy, Myocardial Infarction, Gastroesophageal Reflux, Cholelithiasis, Mesenteric Ischemia, Hemolysis, Splenic Sequestration, Aplastic Crisis, Priapism, Avascular Necrosis, Osteomyelitis, Septic Arthritis, Stroke, Multi-organ failure

Guideline Inclusion Criteria:

All children, adolescents and young adults presenting to Dell Children's Medical Center and Dell Children's North with a history of sickle cell disease and with acute pain episode.

Guideline Exclusion Criteria:

Patients without sickle cell disease presenting with pain.

Diagnostic Evaluation:

Labs: CBC w/diff, Abs Retic, Type & Screen, CMP, UA, Blood Culture (if febrile), other lab studies at provider discretion.

Radiology studies: CXR (2 view) if respiratory symptoms (cough, chest pain, hypoxia, fever, focal exam). Other imaging at provider's discretion.

Clinical Management:

- **Initial Management:** Appropriate Triage, Establish Venous Access, Initiate Pain Management
- **Secondary Management:** Admission to hospital may be necessary as well if pain is uncontrolled, or if the patient is febrile / ill-appearing. Consult Pedi Hematology if the pain is uncontrolled after initial management, the patient is febrile, or admission is deemed necessary.

Outcome Measures:

- Time to IN Fentanyl (if not refused)
- Time to first IV opioid
- Length of Stay in ED/CBCC Heme
- Time to First Pain assessment
- Time between Pain Assessments
- Use of ED SCD Pain Powerplan
- Total number of medications given in ED
- Number of patients discharged from ED/CBCC Heme clinic vs admitted

Pain Management Algorithm for Patients With Sickle Cell Disease in Vaso-Occlusive Crisis

Emergency Department/Outpatient Hematology Clinic

Emergency Level: Triage Level 2
Patients may call CBCC Heme clinic if having pain and may be told to come into the clinic for treatment. If after hours or unable, they will be directed to present to DCMC ED.

PURPOSE:
To administer pain medication within 30 minutes of patient's arrival to the Emergency Department & CBCC Heme Clinic

- Administer **Fentanyl 2 mcg/kg intranasal (max 100 mcg/dose)**
- Place PIV/Access Port
- Obtain labs: CBC, retic, CMP, urine HCG (females >10 years)
  - If ill-appearing: T&S, Hgb electrophoresis (Stat)
- If febrile: Use SCD Fever Pathway concurrently
- If chest pain w/ hypoxis or fever: Concurrent acute chest treatment
  Notify hematology if CXR concerning for acute chest
- If ordered, give opioid premeds (i.e. PO diphenhydramine)
- Offer heat packs to painful sites
- Continuous pulse oximetry

Triage Questions
« History of acute chest
« Last pain crisis
« Current fever, cough, chest pain
« Individualized pain plan

For patients with an individualized pain plan, check for a High Alert Plan (HAP). Contact hematology provider prior to 1st medication.

GOAL:
0-30 Minutes

Administer:

- Morphine 0.1 - 0.2 mg/kg/dose IV (max 8 mg) **OR**
- Hydromorphone 0.015 mg - 0.02 mg/kg/dose IV (max 1 mg) **OR**
- Fentanyl 2 mcg/kg IV (max 100 mcg) - ED ONLY

AND

- Ketorolac [1] IV 0.5 mg/kg/dose IV (<16yo max 15mg, >16yo max 30mg) x 1 dose

AND

- 10ml/kg NS bolus (max 1L) over 60 minutes. If concern for dehydration, give 20ml/kg bolus (max 1L).
- Then start 1xM IVF.

If unable to obtain IV access: Oxycodone 0.1 mg/kg PO (max 10 mg)

Contraindications to ketorolac:
- Pregnancy
- Renal impairment
- Last dose of ketorolac within 5 days
- Last dose ibuprofen within 6 hours
- Bleeding concerns
- History of or concern for renal impairment

1

GOAL:
31-60 Minutes

Pain Improved?

YES

NO

- Give second dose of narcotic pain medication
- Notify provider before administration

! RN to reassess pain 30 minutes after each pain medication administration and notify provider.
If patient asleep, wake patient up to reassess pain as directed.

GOAL:
61-90 Minutes

Discharge Criteria:
- Observe for 1 hour post narcotic
- Encourage PO intake
- If patient comfortable managing pain at home, discharge home with pain plan/regimen
- Follow up appointment in clinic

ED:
- Page Hematology for prior to discharge for follow up plan
- Refill home pain medication for 2-3 day supply if needed

Clinic:
- Ensure opioid refill if needed

Pain Improved?

YES

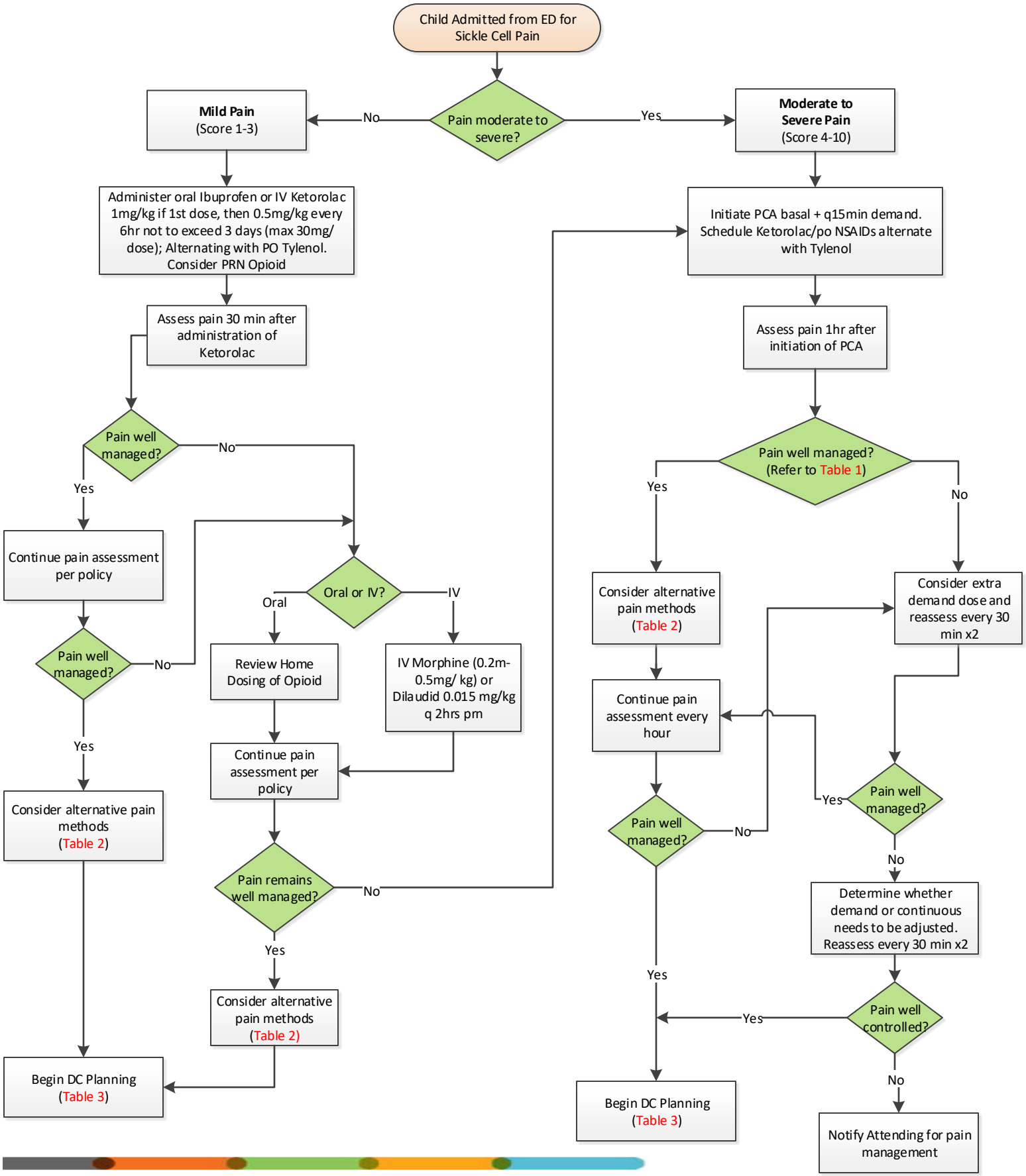
NO

- Give third dose of narcotic pain medication
- **ED:** Contact Hematology Team for further management
- **Clinic:** Provider decision on patient admission to inpatient unit or discharge home with appropriate follow up plan.

Last Updated: April 2023

Sickle Cell Pain Management (Inpatient) Pathway

Evidence-Based Outcomes Center



Last Updated: April 2023

Table 1

Pain – Questions to ask to Evaluate PCA Effectiveness:

- *Is the demand dose helping?*
  - Yes, but it makes me fall asleep every time, but doesn't last the full 15/20 minutes
    →Consider decreasing the bolus dose and interval
  - Yes, but it doesn't last the full 10/15/20 minutes
    →Consider decreasing the dosing interval
  - No, I don't feel it at all
    →Consider increasing the demand dose
- *Do you feel your pain has improved since starting the PCA?*
  - Yes, a little bit, but I'm still hitting my button a lot
    →Consider increasing the continuous +/- demand (depending on demand answers above)
  - Yes, it's helping a lot
    →Continue as is!
  - No, I'm still a 10/10
    →Consider increasing both the continuous and demand (as demand questions above as well)

Table 2

Alternative/Adjuvant Pain Management to consider:

- TENS unit (PT consult)
- Lidocaine patch
- Virtual Reality (Social Work)
- Psychology/Psychiatry consult?
- Simple/Exchange Transfusion
- Prolonged NSAID use
- For those NOT on a PCA, consider PCA with demand only to give some control of their pain management

Table 3

Discharge Planning

- Start Methadone/wean PCA
- Consult Case management 48 hours prior to discharge in case prior authorization/home health needed
- Provider should send prescriptions to DCOP/home pharmacy 48 hours prior to discharge
- Social work- school plan

Methods

Existing External Guidelines/Clinical Pathways

Existing External Guideline/Clinical Pathway	Organization and Author	Last Update
Sickle Cell Patient in Acute Pain Crisis Guideline	Zora Rogers	2012
Guideline: Sickle Cell DIsease (SCD) Patients with Pain	University of Chicago	9/29/2015
Sickle Cell Disease in Vaso-Occlusive Crisis Evidence-Based Guideline	Texas Children's Hospital	July 2017
Vanderbilt Pain Algorithm	Vanderbilt	No Date
ED SCD Pathway	OU Children's	No Date
ED Pathway for Evaluation/Treatment of Children with Sickle Cell Disease and Pain	Children's Hospital of Philadelphia	2020
Sickle Cell Disease	John Hopkins	2022
Sickle Cell Disease (SCD) Pain Crisis Pathway	Arkansas Children's	2019

Any published clinical guidelines have been evaluated for this review using the **AGREE II criteria**. The comparisons of these guidelines are found at the end of this document. **AGREE II criteria** include evaluation of: Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity of Presentation, Applicability, and Editorial Independence.

Review of Relevant Evidence: Search Strategies and Databases Reviewed

Search Strategies	Document Strategies Used
Search Terms Used:	Sickle Cell Disease, Pain Management, SCD
Years Searched - All Questions	1995 - 2023
Language	English
Age of Subjects	0-18 years old
Search Engines	PubMed, Scholar Google
EBP Web Sites	
Professional Organizations	www.sicklecelldisease.org www.hematology.org www.nhlbi.nih.gov www.ampainsoc.org https://www.acep.org/patient-care/sickle-cell/
Joint Commission	
Government/State Agencies	http://www.cdc.gov/ncbddd/sicklecell/index.html

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2023 Revision

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Approved by the Pediatric Evidence-Based Outcomes Center Team

Revision History
Original Date Approved: May 20, 2019
Revision Dates: **April 2023** - Addition of Pain Management Algorithm for Patients With Sickle Cell Disease in Vaso-Occlusive Crisis

Emergency Department/Outpatient Hematology Clinic
Next Review Date: May 2027

Sickle Cell with Pain EBOC Team:
Dory Collette, RN, CCRN
Robert Mignacca, MD
Mark Tabarrok, MD
Molly McNaull, PharmD
Daryl Mozygemba, RN, MSN, CPNP-PC
Amber Bills, MSN, RN, CPN, CPON
Debra Rodriquez, MSN, RN
Denita Lyons, BSN, RN, CPEN
Anne Raines, MSN, RN, CPON
Frank James, MBA
Carmen Garudo, PM

EBOC Committee:
Lynn Thoreson, DO
Sarmistha Hauger, MD
Patty Click, RN
Sheryl Yanger MD
Tory Meyer, MD
Meena Iyer, MD
Nilda Garcia, MD
Amanda Puro, MD
Lynsey Vaughan, MD

2023 Review team:
Amber Bills, MSN, RN, CPN, CPON
Alicia Chang, MD
Katie Bookout, MSN, RN, CPN
Mark Tabarrok, MD
Carmen Garudo, EBOC PM

Recommendations

Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible.

Approval Process

EBOC guidelines are reviewed by DCMC content experts, the EBOC committee, and are subject to a hospital wide review prior to implementation. Recommendations are reviewed and adjusted based on local expertise.