Clinical Pathways

Sickle Cell: Management of Acute Pain Crisis

Taryn J. Hamre, DNP, APRN
William Zempsky, MD
Natalie Bezler, MD
Donna Boruchov, MD

Connecticut Children's

BEST CHILDREN'S HOSPITALS U.S.News RANKED IN 4 SPECIALTIES 2018-19

MAGNET RECOGNIZED AMERICAN NURSES CREDENTIALING CENTER

2019 WOMEN'S CHOICE AWARD® BEST CHILDREN'S HOSPITALS

What is a Clinical Pathway?

An evidence-based guideline that decreases unnecessary variation and helps promote safe, effective, and consistent patient care.

Background/Why is Pathway Necessary?

- Sickle Cell Disease (SCD) is the most common genetic disease in the U.S. It is caused by a mutation in the hemoglobin beta chain in which glutamic acid is substituted with valine. The CDC (2019) estimates that over 100,000 Americans are affected by SCD. The disease can affect multiple organ systems and decrease life expectancy.
- Lack of standardized national guidelines for acute pain management in patients with Sickle Cell Disease
- No standardized approach to inpatient Sickle Cell Acute Pain Management at Connecticut Children's which leads to inconsistency in care
- High readmission rates
- Prolonged IV opioid management
- Opioid Crisis
- Opioid induced hypersensitivity
- National goal to reduce the use of long-acting opioids for patients with non-cancer pain (increased side effects and risks; decreased efficacy)

Objectives of Pathway

- Standardize sickle cell acute pain treatment
- Decrease LOS and readmission rates
- Decrease the time patients receive intravenous opioids
- Improve timely consultation of the Pain Team (if needed)
- Improve timely administration of multi-modal treatments
- Encourage early mobilization
- Increase us of the Acute Pain Admission comprehensive order set

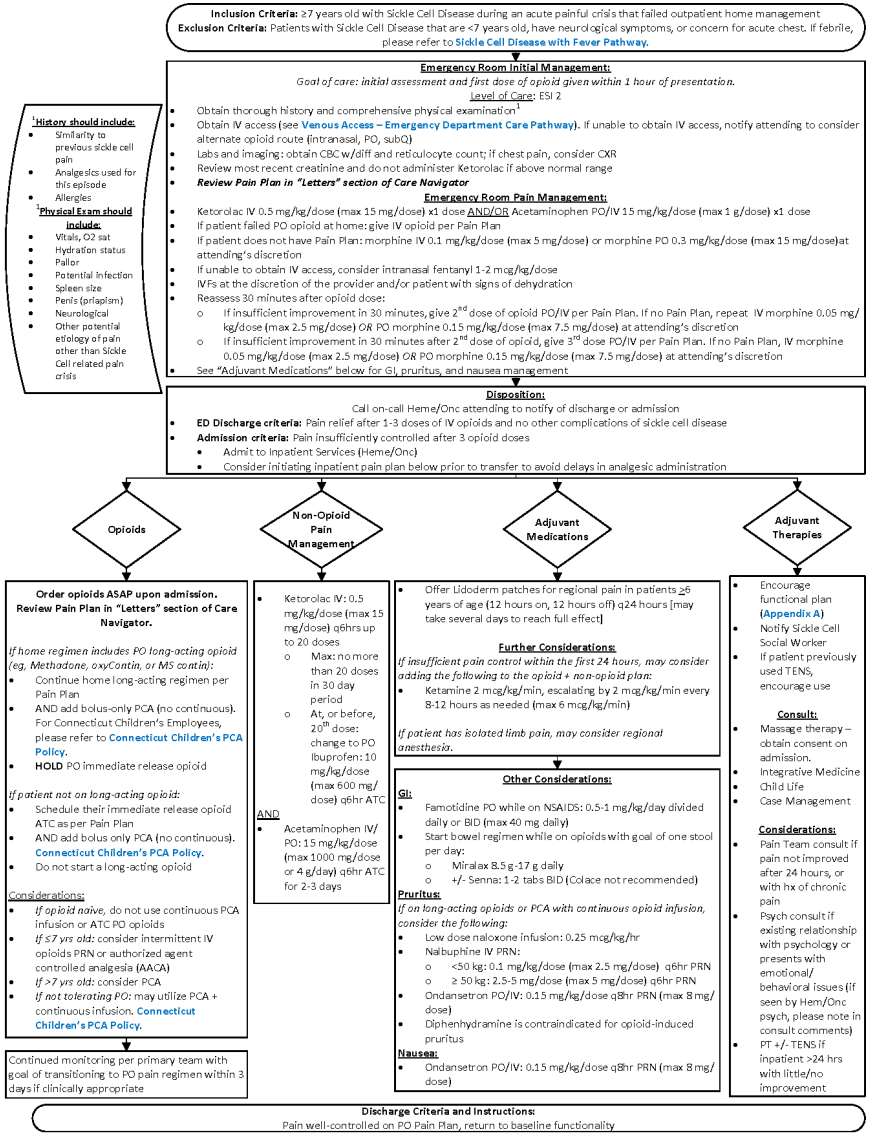
This is the Sickle Cell with Acute Pain Clinical Pathway.

We will be reviewing each component in the following slides.

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.



CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

LAST UPDATED: 04.22.21

- This pathway is for patients >7 years, as most sickle cell patients that are admitted are over the age of 7, as well as the PCA cut off for nurse controlled vs independent control is 7 years.
- Pain Plan – each patient will have a pain plan from Hem/Onc in the "Chart Review" section, under "Letters".
- Reviewing the H&P is critical, not all pain is SCD pain. For example, you would not want to miss an appendicitis.

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL

Inclusion Criteria: ≥7 years old with Sickle Cell Disease during an acute painful crisis that failed outpatient home management
Exclusion Criteria: Patients with Sickle Cell Disease that are <7 years old, have neurological symptoms, or concern for acute chest. If febrile, please refer to **Sickle Cell Disease with Fever Pathway**.

[1]History should include:
- Similarity to previous sickle cell pain
- Analgesics used for this episode
- Allergies

[1]Physical Exam should include:
- Vitals, O2 sat
- Hydration status
- Pallor
- Potential infection
- Spleen size
- Penis (priapism)
- Neurological
- Other potential etiology of pain other than Sickle Cell related pain crisis

Emergency Room Initial Management:

Goal of care: initial assessment and first dose of opioid given within 1 hour of presentation.

Level of Care: ESI 2

- Obtain thorough history and comprehensive physical examination[1]
- Obtain IV access (see **Venous Access – Emergency Department Care Pathway**). If unable to obtain IV access, notify attending to consider alternate opioid route (intranasal, PO, subQ)
- Labs and imaging: obtain CBC w/diff and reticulocyte count; if chest pain, consider CXR
- Review most recent creatinine and do not administer Ketorolac if above normal range
- ***Review Pain Plan in "Letters" section of Care Navigator***

Emergency Room Pain Management:

- Ketorolac IV 0.5 mg/kg/dose (max 15 mg/dose) x1 dose AND/OR Acetaminophen PO/IV 15 mg/kg/dose (max 1 g/dose) x1 dose
- If patient failed PO opioid at home: give IV opioid per Pain Plan
- If patient does not have Pain Plan: morphine IV 0.1 mg/kg/dose (max 5 mg/dose) or morphine PO 0.3 mg/kg/dose (max 15 mg/dose)at attending's discretion
- If unable to obtain IV access, consider intranasal fentanyl 1-2 mcg/kg/dose
- IVFs at the discretion of the provider and/or patient with signs of dehydration
- Reassess 30 minutes after opioid dose:
  - If insufficient improvement in 30 minutes, give 2nd dose of opioid PO/IV per Pain Plan. If no Pain Plan, repeat IV morphine 0.05 mg/kg/dose (max 2.5 mg/dose) *OR* PO morphine 0.15 mg/kg/dose (max 7.5 mg/dose) at attending's discretion
  - If insufficient improvement in 30 minutes after 2nd dose of opioid, give 3rd dose PO/IV per Pain Plan. If no Pain Plan, IV morphine 0.05 mg/kg/dose (max 2.5 mg/dose) *OR* PO morphine 0.15 mg/kg/dose (max 7.5 mg/dose) at attending's discretion
- See "Adjuvant Medications" below for GI, pruritus, and nausea management

Order opioids ASAP upon admission. Review Pain Plan in "Letters" section of Care Navigator.

If home regimen includes PO long-acting opioid (eg, Methadone, oxyContin, or MS contin):
- Continue home long-acting regimen per Pain Plan
- AND add bolus-only PCA (no continuous). For Connecticut Children's Employees, please refer to **Connecticut Children's PCA Policy**.
- **HOLD** PO immediate release opioid

If patient not on long-acting opioid:
- Schedule their immediate release opioid ATC as per Pain Plan
- AND add bolus only PCA (no continuous). **Connecticut Children's PCA Policy.**
- Do not start a long-acting opioid

Considerations:
- *If opioid naive,* do not use continuous PCA infusion or ATC PO opioids
- *If ≤7 yrs old:* consider intermittent IV opioids PRN or authorized agent controlled analgesia (AACA)
- *If >7 yrs old:* consider PCA
- *If not tolerating PO:* may utilize PCA + continuous infusion. **Connecticut Children's PCA Policy.**

Continued monitoring per primary team with goal of transitioning to PO pain regimen within 3 days if clinically appropriate

- Ketorolac IV: 0.5 mg/kg/dose (max 15 mg/dose) q6hrs up to 20 doses
  - Max: no more than 20 doses in 30 day period
  - At, or before, 20th dose: change to PO Ibuprofen: 10 mg/kg/dose (max 600 mg/dose) q6hr ATC

AND

- Acetaminophen IV/PO: 15 mg/kg/dose (max 1000 mg/dose or 4 g/day) q6hr ATC for 2-3 days

- Offer Lidoderm patches for regional pain in patients ≥6 years of age (12 hours on, 12 hours off) q24 hours [may take several days to reach full effect]

Further Considerations:

If insufficient pain control within the first 24 hours, may consider adding the following to the opioid + non-opioid plan:
- Ketamine 2 mcg/kg/min, escalating by 2 mcg/kg/min every 8-12 hours as needed (max 6 mcg/kg/min)

If patient has isolated limb pain, may consider regional anesthesia.

Other Considerations:

GI:
- Famotidine PO while on NSAIDS: 0.5-1 mg/kg/day divided daily or BID (max 40 mg daily)
- Start bowel regimen while on opioids with goal of one stool per day:
  - Miralax 8.5 g-17 g daily
  - +/- Senna: 1-2 tabs BID (Colace not recommended)

Pruritus:
If on long-acting opioids or PCA with continuous opioid infusion, consider the following:
- Low dose naloxone infusion: 0.25 mcg/kg/hr
- Nalbuphine IV PRN:
  - <50 kg: 0.1 mg/kg/dose (max 2.5 mg/dose) q6hr PRN
  - ≥ 50 kg: 2.5-5 mg/dose (max 5 mg/dose) q6hr PRN
- Ondansetron PO/IV: 0.15 mg/kg/dose q8hr PRN (max 8 mg/dose)
- Diphenhydramine is contraindicated for opioid-induced pruritus

Nausea:
- Ondansetron PO/IV: 0.15 mg/kg/dose q8hr PRN (max 8 mg/dose)

- Encourage functional plan (**Appendix A**)
- Notify Sickle Cell Social Worker
- If patient previously used TENS, encourage use

Consult:
- Massage therapy – obtain consent on admission.
- Integrative Medicine
- Child Life
- Case Management

Considerations:
- Pain Team consult if pain not improved after 24 hours, or with hx of chronic pain
- Psych consult if existing relationship with psychology or presents with emotional/behavioral issues (if seen by Hem/Onc psych, please note in consult comments)
- PT +/- TENS if inpatient >24 hrs with little/no improvement

Discharge Criteria and Instructions:
Pain well-controlled on PO Pain Plan, return to baseline functionality

CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

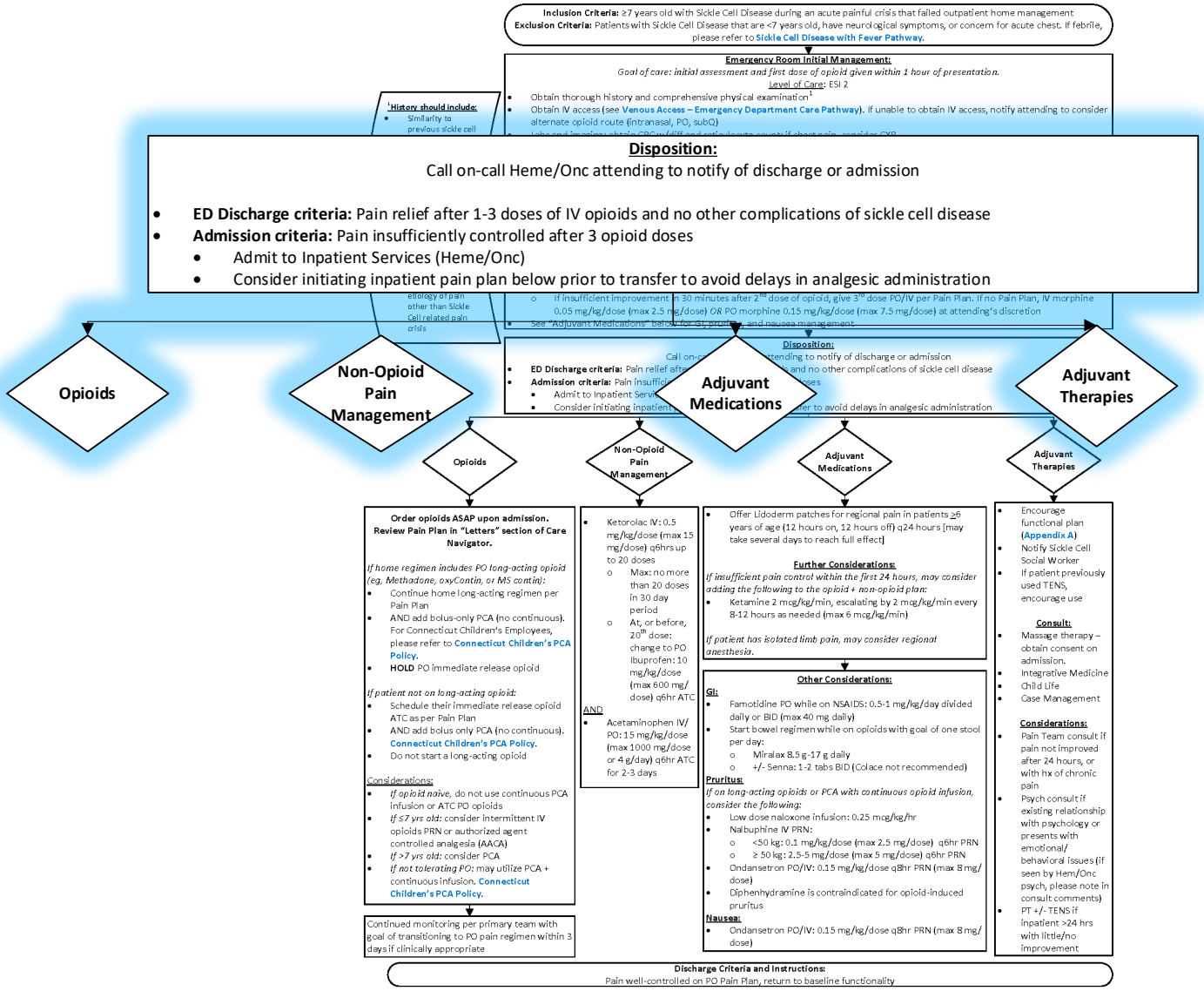
LAST UPDATED: 04.22.21

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.

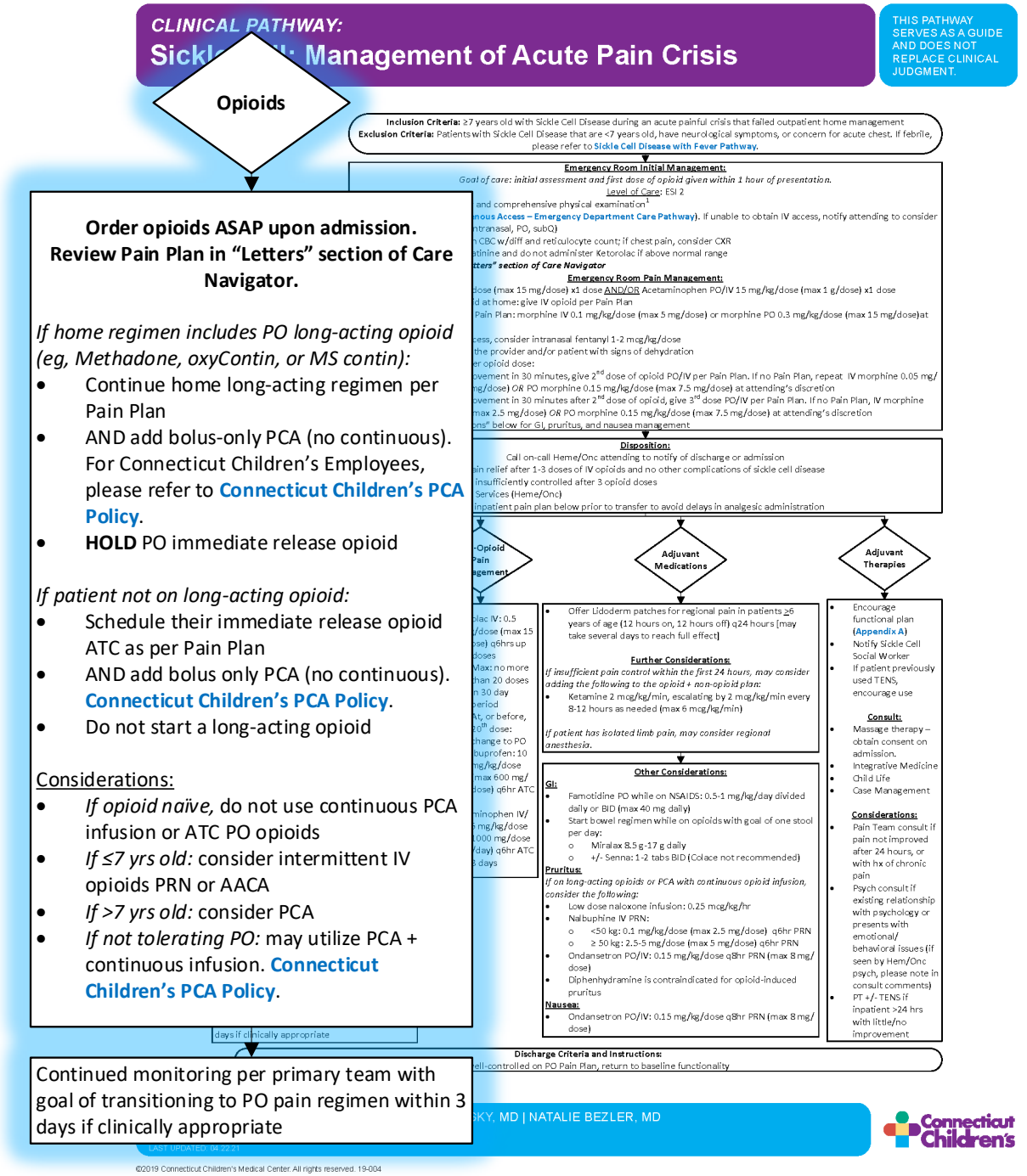
- If pain is still uncontrolled after 3 doses of opioids in the ED, the patient will be admitted.
- Further care will be then broken down into opioids, no-opioids, adjuvant medications, and adjuvant therapies.



CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

LAST UPDATED: 04.22.21

- Opioids should be ordered ASAP upon admission.
- Specific opioid management is based on each patient's Pain Plan.

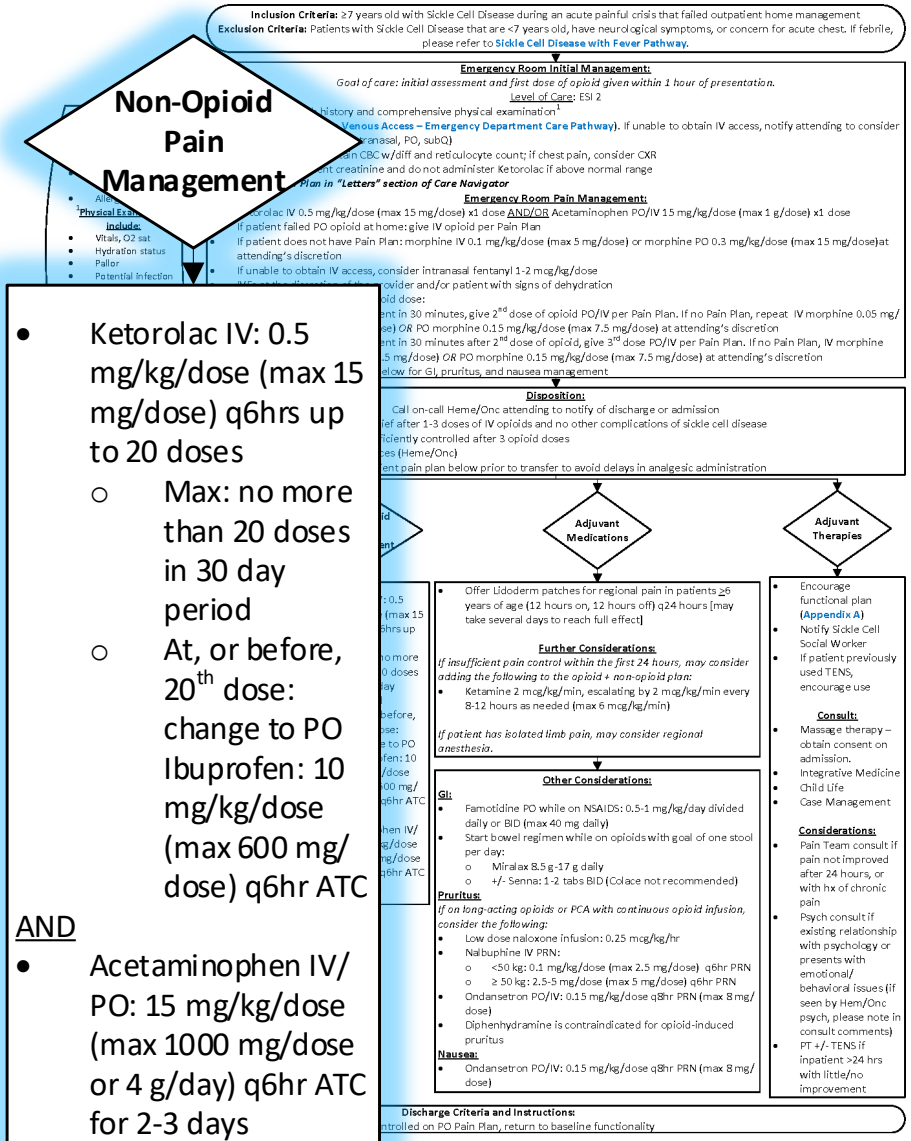


- Non-opioid pain management can include ketorolac and acetaminophen.

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.



Non-Opioid Pain Management

- Ketorolac IV: 0.5 mg/kg/dose (max 15 mg/dose) q6hrs up to 20 doses
  - Max: no more than 20 doses in 30 day period
  - At, or before, 20th dose: change to PO Ibuprofen: 10 mg/kg/dose (max 600 mg/dose) q6hr ATC

AND

- Acetaminophen IV/ PO: 15 mg/kg/dose (max 1000 mg/dose or 4 g/day) q6hr ATC for 2-3 days

CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

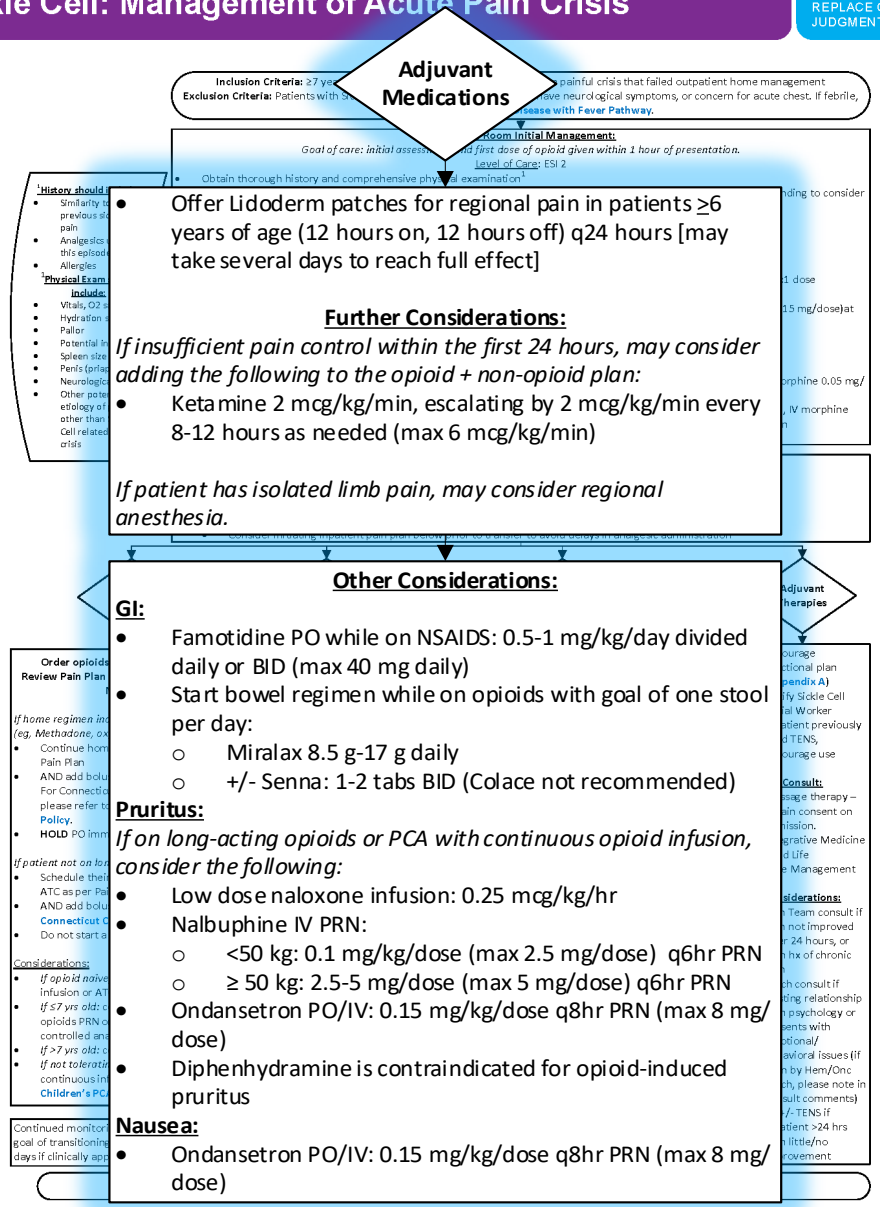
LAST UPDATED: 04.22.21

- Adjuvant medications can help with pain, but also can help with side effects of the medications.
- Considerations should include scheduled lidoderm patches or ketamine for additional pain support.
- GI considerations should include a bowel regimen, ranitidine or famotidine, and/or ondansetron.
- Treatment of pruritus, if present, should also be considered.

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.



CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

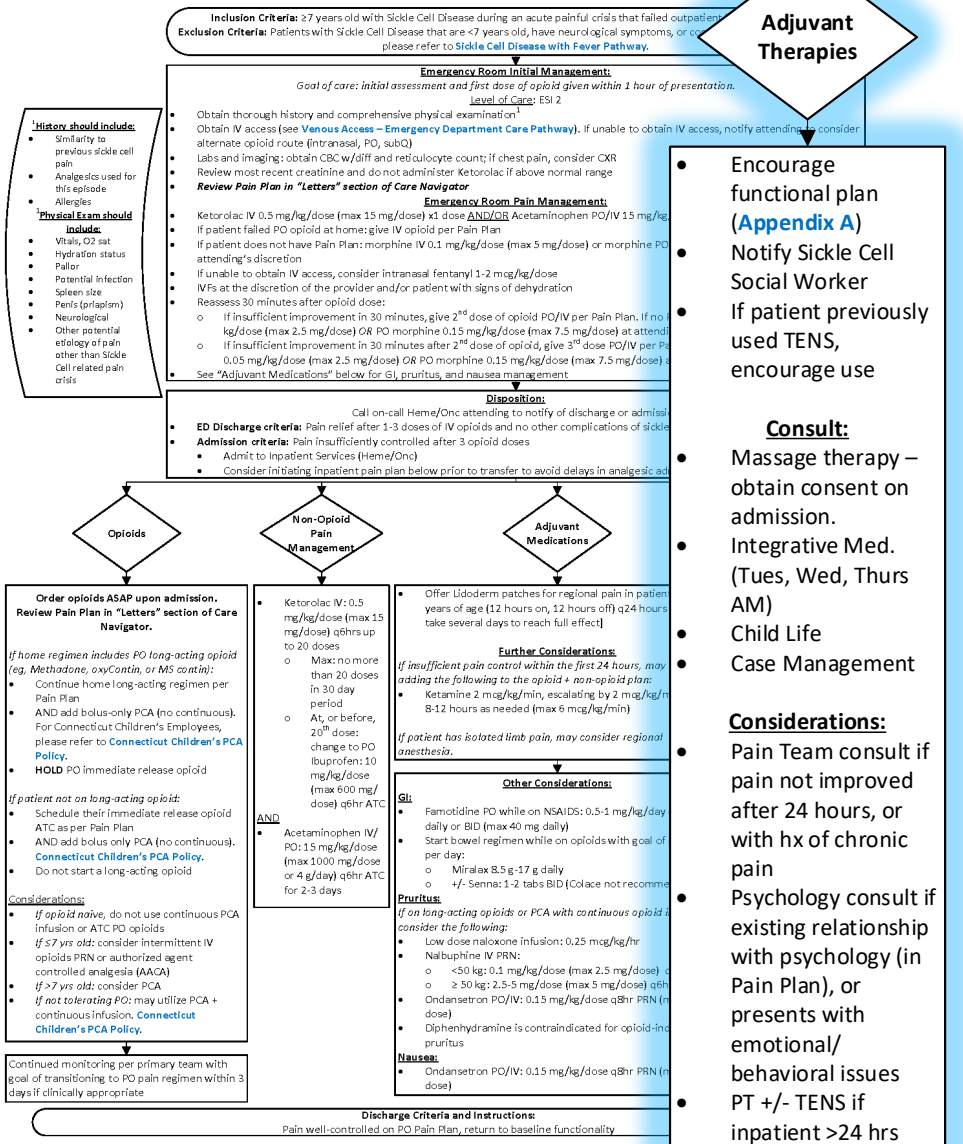
LAST UPDATED: 04.22.21

- All patients should be encouraged to follow a functional plan.
- Consults to be considered include massage therapy, integrative medicine, child life, and case management.
- If the patient has a history of chronic pain, or the patient's pain is not improving after 24 hours, the Pain Team can be consulted.

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.



CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

LAST UPDATED: 04.22.21

Appendix A is the functional plan that should be encouraged early on.

Appendix A: Functional Plan for Patients with Sickle Cell

1.) Regulate sleep/wake cycle - Lights on/blinds open 0900, lights off/electronics off 2200.
2.) Changing for bed into "night clothes" and getting dressed in clothes in AM (if able).
3.) Daily or every other day shower.
4.) Complete activities of daily living (ADL's) independently as tolerated.
5.) Out of bed (OOB) for meals/during meal times if not eating. As admission progresses, OOB more than exclusively for meals (after day one or two) with a rest break in bed in the morning and in the afternoon for up to 1 hour only.
6.) Participation in floor activities. Out of bed, preferably in play room rather than bed side, for special Child Life events, Hole in the Wall Gang Camp activities and art/play projects.
7.) For frequent flyers: school work.
8.) Walks around med/surg unit per PT and/or Primary Team

CONTACTS: TARYN HAMRE, DNP, APRN | BILL ZEMPSKY, MD | NATALIE BEZLER, MD

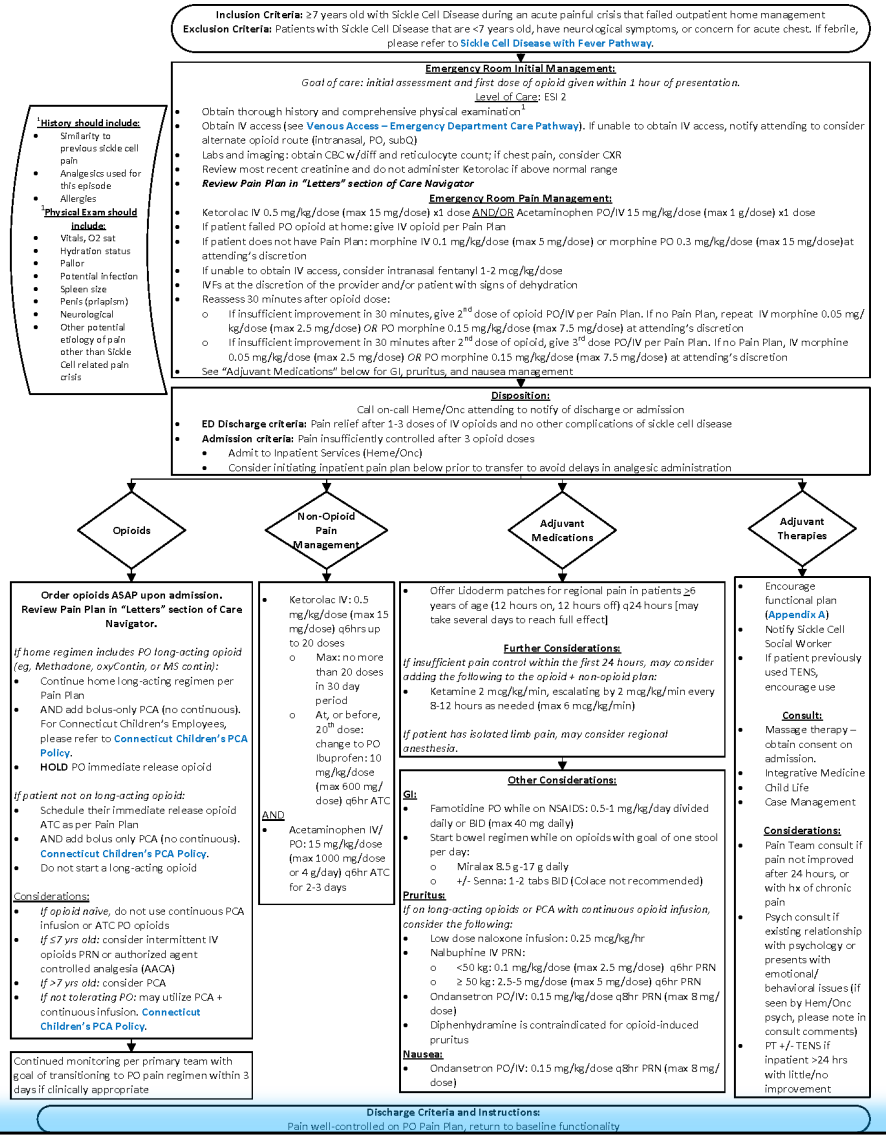
LAST UPDATED: 04.22.21

- Discharge criteria includes pain being well-controlled on a PO pain plan and the patient has returned to their baseline functionality.

CLINICAL PATHWAY:

Sickle Cell: Management of Acute Pain Crisis

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.



Discharge Criteria and Instructions:

Pain well-controlled on PO Pain Plan, return to baseline functionality

Review of Key Points

- Timely assessment and initiation of pain plan is essential
- An interdisciplinary, multimodal approach to acute Sickle Cell Disease pain management is ideal
- Our Pain Team is available to help when needed

Quality Metrics

- Percentage of eligible patients who utilize the pathway order set
- Average time from ED arrival to first opioid administered (oral or IV) in ED (minutes)
- Percentage of ED patients who are admitted
- Average time on IV opioids after arrival to medical-surgical floor (hours)
- Average time from arrival to ED to PCA initiation (minutes)
- Average time from arrival to medical-surgical floor to PCA initiation (minutes)
- Percentage of admitted patients with pain team service consult ≤ 24 hours from admission
- Percentage of admitted patients who have orders for any of the following adjuvant therapies: PT, Psychology, Integrative Medicine, or Massage Therapy
- ALOS (days, IP/OBS) and ALOS (minutes, ED)
- Readmissions within 7 days
- Readmissions within 30 days

Pathway Contacts

- Taryn J. Hamre, DNP, APRN – Division of Pain and Palliative Medicine
- William Zempsky, MD – Division of Pain and Palliative Medicine
- Natalie Bezler, MD – Hematology/Oncology
- Donna Boruchov, MD - Hematology/Oncology

References

- Balsamo L, Shabanova V, Carbonella J, Szondy MV, Kalbfeld K, Thomas DA, Santucci K, Grossman M, Pashankar F. Improving Care for Sickle Cell Pain Crisis Using a Multidisciplinary Approach. *Pediatrics*. 2019 May;143(5):e20182218. doi: 10.1542/peds.2018-2218.
- Brandow AM, Nimmer M, Simmons T, et al. Impact of emergency department care on outcomes of acute pain events in children with sickle cell disease. *Am J Hematol.* 2016 Dec;91(12):1175-1180.
- Brandow AM, Zappia KJ, Stucky CL. Sickle cell disease: a natural model of acute and chronic pain. *Pain*. 2017 Apr;158 Suppl 1(Suppl 1):S79-S84.
- Dampier C, Palermo TM, Darbari DS, Hassell K, Smith W, Zempsky W. AAPT Diagnostic Criteria for Chronic Sickle Cell Disease Pain. *J Pain*. 2017 May;18(5):490-498.
- Field, JJ, Ballas SK, Campbell CM, Crosby, LE, Dampier C, Darbari DS, McClish DK, Smith W, Zempsky WT. AAAPT Diagnostic Criteria for Acute Sickle Cell Disease Pain. *J Pain*. 2019 Jul;20(7):746-759.
- Sheehy KA, Lippold C, Rice AL, Nobrega R, Finkel JC, Quezado ZM. Subanesthetic ketamine for pain management in hospitalized children, adolescents, and young adults: a single-center cohort study. *J Pain Res.* 2017 Apr 5;10:787-795.
- Zempsky WT, Loiselle KA, Corsi JM, Hagstrom, JN. Use of Low-dose Ketamine Infusion for Pediatric Patients With Sickle Cell Disease-related Pain. *Clin J Pain*. 2010 Feb;26(2):163-7.

Additional Resources

- Pain List Serv – feedback from other Children's Hospitals
- Sickle Cell FAB
- Dr Zempsky's participation in national Sickle Cell Meetings to address pain management
- Hospital-wide stakeholder meeting
- Sicklecelldisease.org
- New England Pediatric SC Consortium

Thank You!

About Connecticut Children's Clinical Pathways Program

The Clinical Pathways Program at Connecticut Children's aims to improve the quality of care our patients receive, across both ambulatory and acute care settings. We have implemented a standardized process for clinical pathway development and maintenance to ensure meaningful improvements to patient care as well as systematic continual improvement. Development of a clinical pathway includes a multidisciplinary team, which may include doctors, advanced practitioners, nurses, pharmacists, other specialists, and even patients/families. Each clinical pathway has a flow algorithm, an educational module for end-user education, associated order set(s) in the electronic medical record, and quality metrics that are evaluated regularly to measure the pathway's effectiveness. Additionally, clinical pathways are reviewed annually and updated to ensure alignment with the most up to date evidence. These pathways serve as a guide for providers and do not replace clinical judgment.