

INCLUSION CRITERIA

- Patients with sickle cell disease ≥ 1 year old and 10kg who present with musculoskeletal pain secondary to suspected vaso-occlusive crisis

EXCLUSION CRITERIA

- Trauma
- Focal neurologic findings
- Altered mental status
- Respiratory distress
- Hemodynamic instability
- Toxic appearance

MEASURES

- ED LOS
- % of patients given intranasal fentanyl
- Time from arrival to opioid administration

IMAGING¹

- CXR if cough, shortness of breath, tachypnea, retractions, wheezing, chest pain, pulse ox less than baseline, or abnormal lung examination

DISCHARGE CRITERIA²

- Adequate pain relief
- Tolerating PO
- Baseline oxygenation on room air
- Blood counts at or near baseline
- Absence of other complications

PAIN REASSESSMENTS

- Reassess and document pain scores q20 min after giving IV opioid

Initial Assessment

- Enter CC: sickle cell with pain and document pain score
- Notify ED provider for pain medication order
- If febrile or reported fever at home, also implement SCD Fever Pathway



Do not administer ibuprofen

Intranasal Medication

Give intranasal fentanyl as a **bridge** to IV analgesia within **30 mins** of arrival

Diagnostic Evaluation

- CBC, Reticulocyte count, CMP and HCG
- Draw and hold type and screen if volume sufficient
- Obtain 2 view CXR if concern for acute chest¹
- Consider abdominal x-ray or ultrasound if patient presents with abdominal pain or direct hyperbilirubinemia



Do not delay medication administration. Send serum HCG if patient unable to urinate for POCT HCG

Intravenous Medications

- IV opioid (Refer to historical ED visit(s) and/or patient/parent report to guide opioid choice/dose)
- IV ketorolac (if no ibuprofen within 6 hours and Cr normal within 3 months)
- D5 0.45NSS @ maintenance



Avoid excessive IV fluids or boluses which may precipitate acute chest syndrome



If patient sleeping use FLACC score for pain reassessment

Reassess in 20min

Pain continues?

No

Yes

Call Hematology

- Administer IV opioid, 100% initial dose immediately

Reassess in 20min

Pain continues?

No

Yes

Refer to EMR to determine appropriate oral opioid and administer one dose

- Administer IV opioid, 100% initial dose immediately

Reassess in 20min

Pain continues?

No

Yes

Reassess in 60min

Discharge If Meets Criteria²

- Utilize ED Sickle Cell with Pain Disposition Order Set
- Ensure access to home opioids
- RX 3 days of home oral analgesic regimen
- Instruct to take home NSAID regimen ATC 24 hours then PRN
- F/u with Hematology

If pain continues after **3rd dose** of IV opioid, call Hematology for admission. Ensure PRN dose ordered while awaiting transfer to floor

Medication	Dose	Route	Max Dose
Morphine	0.1 mg/kg	IV	6 mg
Hydromorphone	0.015 mg/kg	IV	1.5 mg
Ketorolac	0.5 mg/kg	IV	30 mg
Fentanyl	2 mcg/kg	Intranasal	100 mcg

Abbreviations			
Abbreviation	Terminology	Abbreviation	Terminology
ATC	Around the Clock	LOS	Length of stay
CBC	Complete Blood Count	mg	Milligrams
CC	Chief complaint	Mins	Minutes
CMP	Complete Metabolic Panel	NSAID	Infectious Disease
Cr	Creatinine	PO	By Mouth
CXR	Chest x-ray	POCT	Point of Care Test
ED	Emergency Department	PRN	As needed
EMR	Electronic Medical Record	Q	Every
FLACC	Face, Legs, Activity, Cry and Consolability Scale	RX	Prescription
F/u	Follow-up	SCD	Sickle Cell Disease
HCG	Human Chorionic Gonadotropin	LOS	Length of stay
IV/IO	Intravenous Intraosseous	CXR	Chest x-ray
kg	Kilograms	CC	Chief complaint

Disclaimer: The clinical pathways are based upon publicly available medical evidence and/or a consensus of medical practitioners at Nemours Children's Health ("Nemours") and are current at the time of publication. These clinical pathways are intended to be a guide for practitioners and may need to be adapted for each specific patient based on the practitioner's professional judgment, consideration of any unique circumstances, the needs of each patient and their family, and/or the availability of various resources at the health care institution where the patient is located.

VERSION MANAGEMENT

- Version 1.0 (10/1/2024): Go Live.
- Version 1.1 (9/8/2025): Adjusted medication table and removed IV medications process step.

Questions about this pathway should be directed to **clinicalpathways@nemours.org**.

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