Clinical Guideline

Sickle Cell Vaso-occlusive Crisis (VOC)

Pediatric Emergency Medicine & Hematology-Oncology

This guideline should not replace clinical judgment.

Inclusion criteria: Age 2-18 years, Diagnosis of Sickle Cell disease, Presenting with VOC

Exclusion Criteria: Hypoxia, Fever, Splenomegaly, Uncharacteristic pain

Child with Simple Vaso-occlusive Episode

For patients >10kg, give intranasal (IN) Fentanyl, 1.5 mcg/kg/dose (max 100 mcg/dose), 2 doses 5 minutes apart

Consider trial of only oral oxycodone FIRST, if no prior opioids within last 12 hours

If pain is severe and anticipate need for IV therapy, proceed with PIV placement

If <10kg or refuses fentanyl

Place PIV and check chart for individualized pain plan (IPP)

To find IPP in Cerner, extend date range, go to: Clinical notes > Other documentation > IPP

Pain improved?

No

Yes

Give oral opioid and call fellow to discuss discharge planning (Labs not absolutely necessary if IV opioids not needed)

Individualized Pain Plan?

Yes

No

As soon as possible:

- Complete full evaluation
- Obtain CBC w/diff, retic, T&S (if concern for severe anemia)
- Manage according to pain plan

Pain significantly improved after 2 IV opioid doses according to pain plan?

Yes

No

In the next 30 minutes:

- Compete full assessment
- Obtain CBC w/diff, retic, T&S (if concern for severe anemia)
- Consider IVF if clinically dehydrated
- Give initial IV morphine at 0.1 mg/kg if already received IN fentanyl or 0.2 mg/kg if not, both max 4 mg & ketorolac 0.5 mg/kg (max 30 mg)

Pain improved?

Yes

No

Give oral opioid and call fellow to discuss discharge planning

In the next 30 minutes:

- Give second dose of IV morphine 0.1mg/kg (max 4mg)
- Re-evaluate in 30 minutes

Pain improved?

Yes

No

Pain score 7 or higher?

Yes

No

- Give third dose of IV morphine 0.1mg/kg (max 4mg)
- Re-evaluate in 30 minutes
- If pain significantly improved, give oral opioid and call fellow to discuss discharge planning

Unless pain significantly improved after 3rd dose

Plan Admission:

- Give third dose of IV narcotic, if not already given
- Call fellow to discuss admission plan
- If pain plan exists, order PCA per recommendations
- If no pain plan, order PCA per dosing chart

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For questions concerning this guideline, contact: chorclinicalguidelines@vcuhealth.org

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Next expected update: September 2022

Upon inpatient admission:

- Use Pediatric Sickle Cell Admission Orders Powerplan
- Continue home medications including Folic Acid and (±) Penicillin
- Continue Hydroxyurea if ANC > 1000 and PLT > 80,000
- Continue Ketorolac 0.5 mg/kg/dose (max dose 30mg) every 6 hours scheduled (after 48 hours, switch to scheduled Ibuprofen)
- Start PCA per Individualized Pain Plan if not started in ED; if no pain plan, use Medication Table below
- Assess patient on arrival to floor – may need opioid bolus while awaiting PCA set up

Dose Adjustment Guidelines:

- If the patient has increased pain scores and is using PCA > 3x/hour, consider giving a bolus dose and increasing basal by 20-25%
- Reassess patient within 1 hour after ANY dose adjustments for sedation and efficacy
- Do not increase basal/PCA dosing more frequently than every 3-4 hours

Side Effect Management:

- Bowel Regimen scheduled: MUSH (Docusate/Miralax) + PUSH (Senna) ± Lactulose as needed
- Itching relief with ORAL Diphenhydramine, Hydroxyzine, or Cetirizine as needed
- Nausea relief with Ondansetron as needed

Other:

- Continuous Pulse Oximetry on all PCA patients for the first 48 hours and with any PCA dose escalation
- IV Fluids should be based on oral intake and clinical hydration status. Goal: achieve & maintain euvolemia.
- If patient is unable to eat or drink, maintenance fluids should be maxed at 1 x maintenance fluid rate.
- **Incentive Spirometry – ensure equipment at bedside and within reach of patient; monitor usage**
- Consider PT consult after 24 hours, if specific movement issue identified
- Up & Ambulate at least 2x per shift (mandatory)
- Labs: CBC with Retic at attending/fellow discretion

Medication table

*Ranges listed indicate starting doses for opioid-naive patients

Medication	Dose	MAX INITIAL DOSE
Oxycodone	</=6 months PO: 0.025-0.05 mg/kg/dose every 4-6 hours >6 months PO: 0.1-0.2 mg/kg/dose every 4-6 hours	PO: 5 mg - 10 mg
Morphine	PO: 0.1-0.3 mg/kg/dose every 3-4 hours IV: 0.1-0.2 mg/kg/dose every 3-4 hours	PO: 15 mg IV: 4 mg
Hydromorphone	PO: 0.03-0.08 mg/kg/dose every 3-4 hours IV: 0.015 mg/kg/dose every 3-4 hours	PO: 2 mg IV: 0.6 mg
Morphine PCA (1st line)	Continuous rate: 0.01-0.03 mg/kg/hour PCA dose: 0.02 mg/kg/every 10 min Clinician bolus: 0.05 mg/kg	
Hydromorphone PCA	Continuous rate: 0.001-0.003 mg/kg/hour PCA dose: 0.002 mg/kg every 10 min Clinician bolus: 0.005 mg/kg	

Sickle Cell Vaso-occlusive Crisis Guideline
Executive Summary

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Approved (November 2019)

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References

Kavanaugh PL, et. al. Improving the management of Vaso-Occlusive Episodes in the Pediatric Emergency Department. Pediatrics. 136:4. October 2015. DOI: 10.1542/peds.2014-3470

Krishnamurti L, Smith-Packard B, Gupta A, Campbell M, Gunawardena S, & Saladino R. (2014). Impact of individualized pain plan on the emergency management of children with sickle cell disease. Pediatr Blood Cancer, 61. https://doi.org/10.1002/pbc.25024

Schefft, MR, Swaffar C, Newlin J, Noda C, & Sisler I. (2018). A novel approach to reducing admissions for children with sickle cell disease in pain crisis through individualization and standardization in the emergency department. Pediatric Blood & Cancer, e27274. https://doi.org/10.1002/pbc.27274

PB, L, HP, S & BL, L. (2014). Sickle cell disease in the emergency department. Emergency Medicine Clinics of North America, 32(3), 629–647.

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Retrieval website: **http://www.chrichmond.org/clinicalguideline-sicklecellVOC**

Example:
Children's Hospital of Richmond at VCU, Schefft M, Silverman J, Sisler I, Noda C, Bullock A, Kirshenbaum C, Hanson C. Sickle Cell VOC Guideline. Available from: http://www.chrichmond.org/clinicalguideline-sicklecellVOC