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 IN Fentanyl
If pain >5 on presentation, proceed with PIV placement immediately after IN Fentanyl administration

If <10kg or refuses fentanyl

Pain improved?

No

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

Yes

Give oral opioid and call fellow to discuss discharge planning

Individualized Pain Plan?

Yes

In the next 30 minutes:
- 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
- Consider IVF if clinically dehydrated
- Give initial IV Morphine 0.2mg/kg (max 4mg) & Ketorolac 0.5mg/kg (max 30m)

Pain improved?

Yes

Give oral opioid and call fellow to discuss discharge planning

No

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

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

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

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

Last updated: August 2018
Next expected update: August 2021

On the Floor:

- 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 in 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 (August 2018)

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References

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, Bullock A, Noda C, Kirshenbaum C, Hanson C. Sickle Cell VOC Guideline. Available from: http://www.chrichmond.org/clinicalguideline-sicklecellVOC