


Clinical Guideline

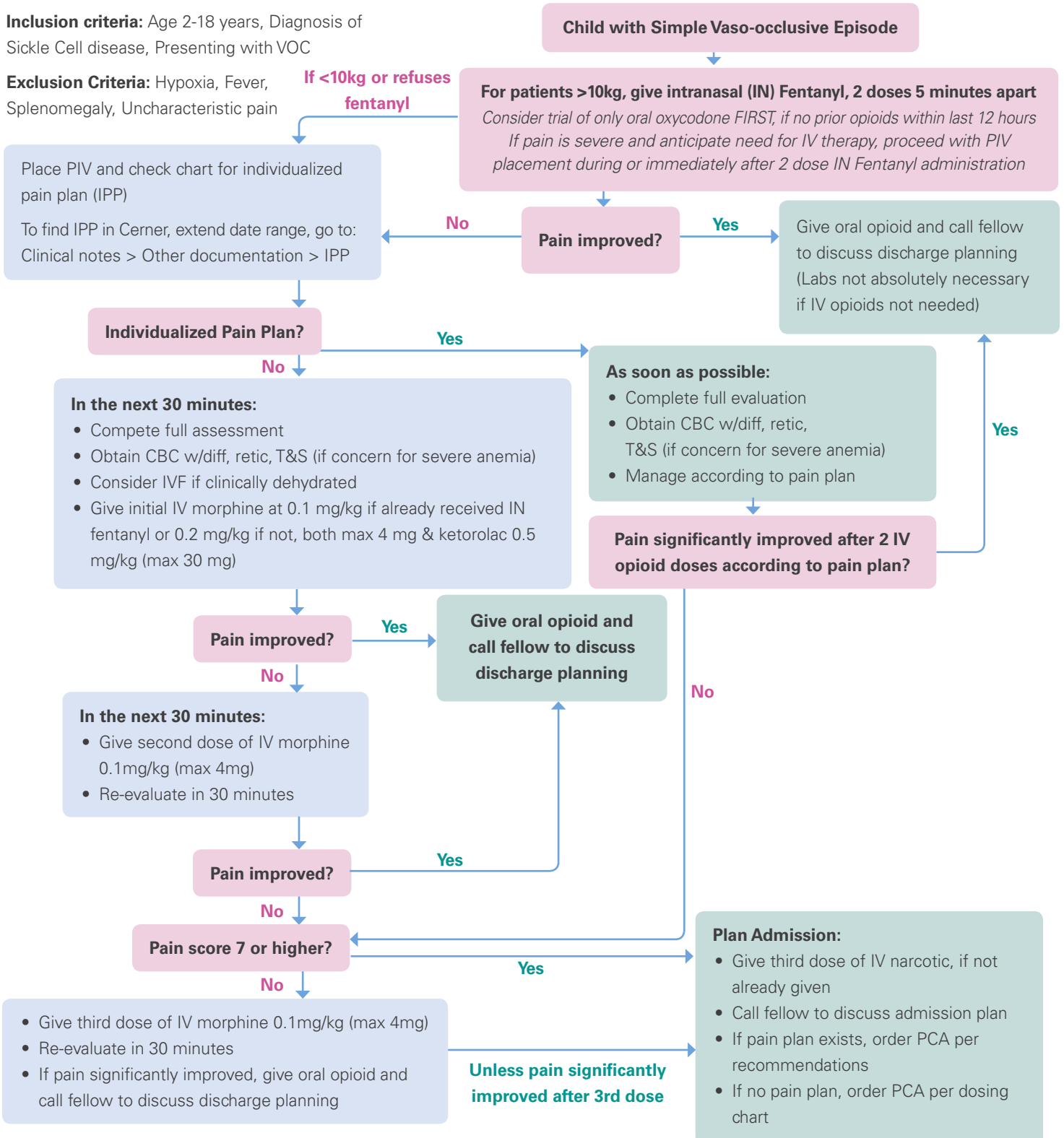
Sickle Cell Vaso-occlusive Crisis (VOC)

 This guideline should not replace clinical judgment.

Pediatric Emergency Medicine & Hematology-Oncology

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

Exclusion Criteria: Hypoxia, Fever, Splenomegaly, Uncharacteristic pain



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 euvoolemia.
- 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

Children's Hospital of Richmond at VCU Sickle Cell VOC Workgroup

Pediatric Hematology-Oncology Owner: Matt Schefft, MD

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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.

Citation

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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>