

SICKLE CELL DISEASE IN VASO-OCCLUSIVE PAIN

Definition: a process in which sickled red blood cells cause tissue ischemia, most often to the bone, resulting in severe pain.

Inclusion Criteria

- Age 1 – 18 years with SCD presenting with pain

Exclusion Criteria

- SCD patient with SCD complications
- Clinical suspicion for ACS in patient

Signs of Critical Complication

- AMS
- New seizure
- Focal neuro deficit
- Fever
- Hypoxia
- Shortness of breath
- Respiratory distress
- Severe headache

Triage and Initial Evaluation

- Vital signs
- Place on pulse ox & cardiac monitoring
- Pain score [FACES see page 2]
- Document last medication time/dose
- Urine HCG all female ≥ 12 years old
- Supplemental oxygen only for SpO2 <95%
- If hypoxia is acute, consider ACS → off algorithm

OFF ALGORITHM

MILD PAIN SCORE 1-3

MODERATE- SEVERE PAIN SCORE > 4

Home opioids used w/in last 12hrs?

Yes

- IV access
- Labs: CBC with diff, reticulocyte count, CMP
- IV fluids 10-20 ml/kg bolus (max 1 L) if dehydrated*
- IV **Morphine dose #1**
dose: 0.1 – 0.2 mg/kg (naïve vs opioid tolerant patient)
- If no NSAID w/in last 6hr → Ketorolac
- PIV access difficult → IN Fentanyl 1mcg/kg (max 100mcg)

No

- No IV access
- Oral Morphine
- Oral fluids
- Warm packs
- Ibuprofen: 10mg/kg (if not used w/in 6h)

Pain well controlled?

No

- IV access
- CBC, retic, CMP

- If pain is not well controlled
- **Morphine dose #2** 0.1- 0.2 mg/kg

30 mins

Yes

- If pain is not well controlled
- **Morphine dose #3** 0.1- 0.2 mg/kg

30 mins

- PO fluid challenge
- ED observation x60 min

Yes

Pain well controlled?

No

- Discuss patient with on-call Peds Hematology Attending
- Admit on PCA (see page 2)

Discharge*

Admit

*** DISCHARGE INSTRUCTIONS**

- Meds: TBD with pediatric heme/onc department
- Follow up: Contact pediatric heme/onc clinic within 24 hours for follow-up
- Return precautions: Inability to control pain at home, inability to tolerate PO, fever, shortness of breath, change in mental status

PEDIATRIC PAIN ASSESSMENT TOOL:

Wong-Baker FACES® Pain Rating Scale



Other Considerations

- ☐ Analgesia
 - Opioids are first line for acute vaso-occlusive pain.
 - Dose should be determined by patient history, review of the medical record or individualized care plan when possible.
 - For patients who are unable to give reliable medication history or have no record available, use established weight-based dosages.
 - If there is significant delay in obtaining IV access, SQ medications should be given to achieve **time to first analgesia within 30 min.**
- ☐ Monitoring
 - Cardiac and pulse oximetry monitoring is strongly encouraged.
 - You may also consider end tidal CO₂.
- ☐ Oxygen
 - Supplemental oxygen only for SpO₂ <95%.
 - If hypoxia is acute, consider ACS or other pathology.
- ☐ IV Fluids
 - IV fluids 10-20 ml/kg max 1L, PRN dehydration/hypovolemia (unless ACS suspected)
- ☐ Labs
 - There are no laboratory values to determine if someone is experiencing acute vaso-occlusive pain
 - CBC with diff, reticulocyte count, CMP is recommended for patients with moderate/severe pain.
- ☐ NSAIDs
 - There is lack of clear evidence to support the use of Ketorolac for the treatment of acute vaso-occlusive pain.
- ☐ Antihistamines
 - Avoid co-administration of IV diphenhydramine and/or promethazine with intravenous opioids due to risk of respiratory depression and oversedation
 - Use non-sedating antihistamines (cetirizine, loratadine, fexofenadine)
 - If needed use PO diphenhydramine for urticaria, pruritis.
- ☐ Hematology consult
 - Discuss pertinent labs
 - Confirm disposition
 - Determine plan for continued scheduled IV opiates or PCA for admitted patients
- ☐ PCA instructions
UNDER CONSTRUCTION

Medication	Route	Dose	Maximum Dose	Frequency	Notes
Ketorolac	IV	0.5 mg/kg	30 mg	- Limited to one dose in the ED - Use is not to exceed 5 consecutive days	Do not give to patients who are pregnant, actively bleeding or have renal dysfunction, <6months of age
Acetaminophen	PO PR	15 mg/kg q6h 30 mg/kg q6h	1000 mg dose 4,000 mg/day		
Fentanyl	IN	1 mcg/kg	100 mcg or 1 ml per nostril	- Limited to two doses - May repeat x1 after 10 min	First-line for pain >4 with if delay in obtaining IV access
Morphine	IV	0.1 mg/kg opioid naive 0.2 mg/kg opioid tolerant	8 mg 10 mg	Repeat as needed q15-30 min. until pain controlled	
Morphine	PO	0.2 mg/kg opiate naive 0.5 mg/kg opioid tolerant		Limited to one oral dose in the ED	
Hydromorphone	IV	0.015 mg/kg	1.5 mg (≤12 yrs) 4 mg (>12 yrs)	Repeat as needed q15-30 min. until pain controlled	

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Created by: Karly Lebherz, M.D.; Anna Suessman, D.O.

Reviewed by: Corey Falcon, M.D.; James Martin, M.D.; Lauren Mutter, M.D.