Assessing and Treating Acute Pain in Children with Sickle Cell Disease

Based on a Canadian Paediatric Society Position Statement

1. **Conduct a comprehensive pain assessment, if possible using self-evaluation of pain to avoid bias[1], considering the unique needs and circumstances for each child/youth. Establish the cause of pain.**
   Evaluate intensity, location, duration, type, impact on function and social factors.
   ✔ Remember, children/youth with sickle cell disease (SCD) can have pain unrelated to SCD. Vaso-occlusive episodes (VOE) cause most, but not all, acute pain episodes.
   ✔ Consider SCD complications[2], which require specific and urgent interventions (See Table 1).

Table 1: Complications of SCD

Presentation	Complications
Thoracic pain	Acute Chest Syndrome (ACS), pulmonary embolus
Acute abdominal pain	Splenic sequestration, cholecystitis, other causes of acute abdomen
Articular pain	Avascular necrosis, osteomyelitis, septic arthritis
Severe headache	Ischemic or haemorrhagic stroke, cerebral sinus venous thrombosis, subdural hemorrhage
Painful erection	Priapism

2. **Develop an acute pain care plan using multimodal, timely, and effective strategies, while preventing further pain and SCD complications.**

A. Physical Pain Management Strategies

- Offer warm packs, warm blankets, caregiver massage of affected area, comfortable positioning of painful areas.
- ✗ ***Avoid cold packs - cold may trigger VOEs in patients with SCD.***

B. Psychological Pain Management Strategies

- Ask the patient and family about their preferences and effective pain management strategies. E.g., consider offering guided relaxation and breathing exercises[3].
- Offer age-appropriate distraction tools (preferably chosen by the child/youth) e.g., music, bubbles, games, puzzles, video games, virtual reality.
- Consider referral to Child Life Specialist or other mental health provider if available.

C. Pharmacological Pain Management Strategies

- Document medication history. Identify what medications:
  - have been taken;
  - usually work best for the patient.
- Create priority care pathways to expedite analgesic administration:
  - Analgesia initiation within 30 to 60 minutes of child/youth's arrival;
  - Rapid analgesic titration if unsatisfactory response.

✔ Administer first-line medications (if no contraindications):
- Acetaminophen around the clock (favour oral or intravenous (IV) route over rectal);
- Nonsteroidal anti-inflammatory drugs (NSAIDs) around the clock (oral or IV).

✔ For moderate to severe pain, rapidly co-administer an opioid per recommended dosing guidelines. See table 2.[4]
- On arrival: intranasal fentanyl (easy administration, rapid onset) followed by oral morphine;
- If repeat doses of morphine needed: Establish IV and provide IV morphine;
- Consider early Patient-Controlled Analgesia;
- Give PEG 3350 if opioids are continued.

✔ Prevent exacerbation of pain and further SCD complications:
- Optimize hydration (preferably orally, IV if needed).

✗ ***Over-hydration increases ACS risk: total fluid intake (oral plus IV) should not exceed a maintenance rate.***

- Prevent ACS and expedite recovery by promoting incentive spirometry and early mobilization.

✗ ***DO NOT transfuse for an uncomplicated pain episode. If transfusion should be considered, always contact the treating hematologist for advice and guidance.***

Table 2: Recommended doses[4]

Medication	Dosing	Daily dose limit	Notes
Acetaminophen by mouth	10 to 15 mg/kg/dose by mouth every 4-6 h (maximum 650 mg to 1 g/dose)	75 mg/kg/day or 4 g/day (whichever is less)	• Administer "round the clock". • Contraindicated in severe hepatic impairment.
NSAID by mouth *Ibuprofen or Naproxen*	Ibuprofen: 10 mg/kg/dose by mouth every 6-8 h (maximum 400 mg to 600 mg/dose) Naproxen: 5 to 7 mg/kg/dose by mouth every 8 to 12 h (maximum 250 mg to 500 mg/dose)	Ibuprofen: 40 mg/kg/day or 2400 mg/day Naproxen: 1 g/day	• Administer "round the clock". • Contraindicated with impaired renal function or GI bleeding. • Choose oral or IV NSAID, ***not both.***
NSAID intravenous *Ketorolac*	0.5 mg/kg/dose IV every 6-8 h (maximum <16 years: 15 mg/dose, ≥16 years: 30 mg/dose)		• Administer "round the clock". • Contraindicated with impaired renal function or GI bleeding. • Limit therapy to 48 h. • Choose oral or IV NSAID, ***not both.***
Fentanyl intranasal	1 to 2 mcg/kg/dose intranasal (maximum 100 mcg/dose)	Use x 1 to 2 doses (maximum 100 mcg total) until alternative mode of analgesia is administered. Use fentanyl 50 mcg/ml for a maximum of 1 ml/nostril.	• Use for patients above 1 year of age. • Divide dose between both nostrils to maximize absorption.
Morphine by mouth	0.2 to 0.5 mg/kg/dose by mouth every 4-6 h (maximum 15 mg/dose)		• Start at lower end in opioid-naïve patients.
Morphine intravenous (intermittent dosing)	0.1 mg/kg/dose IV over 5 minutes, repeat up to every 3 h (maximum 7.5 mg/dose)	May add 0.05 mg/kg (maximum 5 mg) hourly as needed	• Consider lower doses in opioid-naïve patients. • If pain is insufficiently controlled with intermittent morphine dosing, consider initiating a morphine infusion or patient-controlled analgesia (PCA). Consultation with paediatric hematology is recommended. • If patient is intolerant or allergic to morphine, liaise with paediatric hematology for alternative analgesia options.

Canadian Paediatric Society / Société canadienne de pédiatrie
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Consider admission if...
✔ Pain remains significant despite oral analgesia in combination with physical and psychological strategies.
✔ Alternate diagnoses or SCD complication (see Table 1) are suspected.

Consider discharge home if...
✔ Pain is adequately treated with oral analgesics and non-pharmacological strategies.
✔ SCD complications are not suspected.

Time for the patient to go home? Don't forget...

Medical Prescriptions

✔ Prescribe appropriate weight-based doses of acetaminophen and NSAID to optimize analgesia and avoid adverse events.
✗ ***Do not rely on over-the-counter analgesics dosing.***
✔ Ensure patients have sufficient opioids (e.g., 3-5 days; 15-30 doses).
✗ ***Avoid prescribing opioids for > 1 week.***
✔ Consider prescribing a naloxone kit.

Education

Instruct families on the importance of adequately managing pain.
✔ Discuss importance of combining analgesics with physical and psychological strategies.
✔ Discuss importance of round the clock administration of acetaminophen and NSAID, in combination with opioids as needed.
✔ Discuss the potential benefits and harms of opioids as a co-therapy in the context of shared decision making. Taking an opioid for moderate to severe pain improves pain control. For uncontrolled pain, splinting and decreased respiratory effort may result in further complications like acute chest and chronic pain syndromes.

Safety recommendations for opioid use:
✔ Safe administration, tapering, and discontinuation;
✔ Potential side effects the child/youth may experience and how to manage them, including signs of overdose and oversedation;
✔ Safe storage of opioids at home;
✔ Safe disposal of expired or unused medication at the pharmacy;
✔ Symptoms of opioid use disorder.

Follow-up plans

✔ Instruct families to follow-up if pain remains significant despite implementing multimodal pain management strategies or they experience significant opioid side effects.
✔ Ensure follow-up with patient's health care providers (e.g., haematology or paediatric team) within 48h (in person, virtual, by telephone).

Scan here!
For additional information, tools, and to provide feedback on the effectiveness of this resource

linktr.ee/youthinpain

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[1]To learn more about pain evaluation in children:
https://cps.ca/en/documents/position/pain-assessment-and-management
[2]See CPS Position Statement on acute complications of SCD:
https://cps.ca/en/documents/position/acute-complications-with-sickle-cell
[3]To learn about the Comfortability Program for Sickle Cell Pain:
https://www.thecomfortability.com/pages/the-comfort-ability-program-for-sickle-cell-pain
[4]For recommended opioid doses, see:
https://cps.ca/en/documents/position/acute-complications-with-sickle-cell, Table 2

To learn more about SCD, See complete recommendations from CanHaem (The Canadian Haemoglobinopathy Association): https://www.canhaem.org/healthcare-professionals/ *See: Sickle Cell Disease Consensus Statement*

Further information from Health Quality Ontario:
https://www.hqontario.ca/Evidence-to-Improve-Care/Quality-Standards/View-all-Quality-Standards/sickle-cell-disease/The-Quality-Standard-In-Brief

The pain management guidance provided in this document aligns with that outlined in the Pediatric Pain Management Standard:
EN: https://store.healthstandards.org/products/pediatric-pain-management-can-hso-13200-2023-e
FR: https://store.healthstandards.org/products/gestion-de-la-douleur-pediatrique-can-hso-13200-2023-f

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