



INTERPROFESSIONAL PROTOCOL - MUHC

Medication included

No Medication included

THIS IS NOT A MEDICAL ORDER

Title:	Management of Acute Pain in the Pediatric Patient with Sickle Cell Disease
Classification number:	
This interprofessional protocol is attached to:	MUHC Pediatric Opioid Therapy Guidelines Medication Administration Protocol for Ketorolac Medication Administration Protocol Hydromorphone Medication Administration Policy Morphine

INTRODUCTION

- Sickle cell disease (SCD) is an inherited disorder affecting the β -globin subunit of the hemoglobin molecule. When deoxygenated, the altered hemoglobin molecule forms polymers that distort the red blood cell into a sickled shape. Sickled red blood cells have short lifespans, are inflexible and have prolonged transit times through capillary vessels. Micro-vascular occlusion by sickle-shaped red blood cells leads to local hypoxia, ischemia, and eventually tissue damage. Tissue damage generates a variety of inflammatory mediators that activate or sensitize afferent nerve fibres and posterior horn cells of the spinal cord, leading to pain.
- Sickle cell pain is extremely variable and unpredictable in cause, timing, location, and intensity. It may be a manifestation of vaso-occlusion, but may also be a symptom of another process, such as infection. **Pain is an indicator and must be investigated.**
- Sickle cell pain may occur in any part of the body that contains pain receptors, and may involve single or multiple body parts. It can begin or end suddenly or gradually, and severity can range from mild to extremely intense. Vaso-occlusive crises can be precipitated by factors such as exposure to cold, dehydration, strenuous exercise, fatigue, and stress.
- Because patients with SCD may live with daily pain, their pain behaviors may differ from the behaviors health-care practitioners associate with acute pain. It is important to approach pain assessment with as few expectations as possible.

1. PURPOSE

- The following protocol was written with the goal of providing a guideline for the effective and rapid management of acute pain in pediatric patients with sickle cell disease.

2. PROFESSIONALS AND PATIENT POPULATION

Professionals: Physicians, Nurses and Nurses Assistants assuming care for a patient in acute pain due to sickle cell disease, in particular those working in:

- Emergency Department (ED)
- Pediatrics
- Hematology / Oncology
- Anesthesia / Acute Pain Service (APS)

Patient population: Pediatric patients with SCD who are presenting with acute pain

3. ELEMENTS OF CLINICAL ACTIVITY

Professionals are responsible to know the limits and extent of their practice as related to the particular protocol.

IN THE EMERGENCY DEPARTMENT (ED)

All medication below must be prescribed

a. Evaluation of Pain in the ED

- Evaluation of pain according to the MUHC Interprofessional Protocol: Assessment and Management of Pain in the Pediatric Patient.
- Most frequent patients to ED have a resume sheet of last admission and home pain treatment available in the special patient binder.

b. Establishing Analgesia in the ED

****NOTE:** Treatment of pain in the ED should be initiated as soon as possible according to the Canadian Triage and Acuity Scale category. It should not be delayed because of difficult intravenous access or pending lab results. **If difficult IV access is an issue, consider other opioids with alternative routes (for example, oral-transmucosal or intranasal fentanyl, PR morphine and diclofenac, or SC or IM injections of morphine).** **

The **WHO analgesic ladder** is used to direct pharmacological therapy (**Appendix I**). Opioids, in combination with acetaminophen, non-steroidal anti-inflammatory drugs (NSAIDs), and other adjuvants are the mainstay of treatment for pain in sickle cell disease.

- **Acetaminophen:** 15 mg/kg PO/PR Q6h or 12.5 mg/kg PO/PR regular Q4h to a maximum of 1gram/dose. To be started for all patients (unless contraindicated) once a minimum of four hours has passed since the patient's last home dose.
- **NSAIDs:** Patients should be carefully assessed for contraindications to NSAIDs (such as history of GI bleeding or ulcers, renal dysfunction or single kidney). NSAIDs should be started in all eligible patients once a minimum of six hours has passed since patient's last home dose.
 - **Ibuprofen:** 10 mg/kg PO Q6H regular, to a maximum of 400 mg/dose. (above 6 months of age only)
OR
 - **Ketorolac:** 0.5 mg/kg IV x 1 then 0.3-0.5 mg/kg IV Q6H regularly, to a maximum of 30 mg/dose (*for more information, refer to the Medication Administration Protocol for Ketorolac*). (above 1 year old only)
OR
 - **Diclofenac:** 1mg/kg PR every 8 hours, Maximum 150 mg/day. (Suppositories of 12.5mg, 25mg AND 50mg).
- **Opioids:**
 - Patients presenting with severe pain will most likely require IV opioid titration. (**Refer directly to Appendix II page 10, Rapid Morphine titration algorithm.**)
 - **First line opioid; Morphine**, unless contraindications are present. Refer to the Medication Administration Policy Morphine for more details.
 - **Second line only; Hydromorphone.** Risks of oversedation and respiratory depression are increased with this medication. If a patient is receiving it for the first time, a consult to APS is strongly recommended. Refer to the Medication Administration Protocol Hydromorphone for more details.
 - Refer to the **MUHC Pediatric Opioid Therapy Guidelines for monitoring, sedation, and pain scales.**
- **Adjuvants:** Adjuvants such as ketamine and clonidine are often used to treat the pain associated with SCD. With the exception of ketamine in PCA therapy, these are generally not started in the ED.

c. Maintenance of Analgesia in the ED

- Acetaminophen and an NSAID should be given regularly as indicated above.
- If the patient has received his/her first dose of oral opioid in the ED and obtains adequate relief, continue dosing q4h PRN
- If the patient does not obtain adequate relief after receiving his/her first dose of oral opioid in the ED, begin rapid titration of an intravenous opioid (see Appendix II. Rapid Morphine Titration Algorithm for Sickle Cell Patients).

Patient Controlled Analgesia (PCA) in the ED

For safety reasons, only patients who have used PCA in the past will be considered eligible to use PCA in the ED.

- During/after titration, between the hours of 8h00 and 16h00, call APS via locating to assess the possibility to start PCA pump for patient (consult required). From 16h00 to 22h00, contact Anesthesia on call to assess the feasibility to start PCA pump. **No PCA pump therapy will be started at night.** Continue IV bolus/titration until PCA pump is installed. Refer to APS policies for further details on the use of PCA therapy.

d. Non-pharmacological Approaches

- Pharmacological treatments for sickle cell pain should be complemented by psychological, behavioral, and physical modalities. The involvement of parents and child life workers is strongly encouraged. The following are examples of non-pharmacological approaches that may be helpful in managing acute sickle cell pain.
- **Warmth:** While patients with SCD should be counseled to avoid extremes of heat and cold, applying warmth to painful areas during a vaso-occlusive crisis has been reported to help decrease pain. The mechanisms are not clearly understood but are believed to occur via suppression of the transmission of painful stimuli via the A α and C fibers, and local vasodilation.

Warmth can take the form, for example, heated "Magic Bags". The APS has a stock of "Magic Bags" for this purpose, which can be obtained by paging APS (between 8h00-16h00 on weekdays).
- **Guided Imagery:** Guided imagery is a guided distraction technique in which a coach helps the child imagine a pleasant visual image by describing sights, sounds, and physical sensations. At best, the coach should be a trained professional (eg. APS nurses, child life), but parents can be taught to coach their children as well.
- **Hypnosis:** Hypnosis is a cognitive activity where suggestions are used to modify perception. It can also be useful in helping a child gain some degree of control over his or her perception of pain, but requires practice with a trained individual. Some of the staff of the APS are trained in guiding young patients through hypnosis, and may use this technique with those who are eligible.
- **Distraction:** Distraction can help shift a child's attention away from pain, and thus affects the perception of pain. Distraction can involve allowing a child to play a favourite videogame, watch a movie, listen to music, or talk about something non-pain/illness related.

e. Treatment/prevention of opioid-related side-effects:

- **Nausea/vomiting:** **The least sedating agent should always be preferred.**
 - **The first-line choice** is Ondansetron 0.1 mg/kg IV q6h PRN, up to a maximum of 4 mg per dose.
 - **Second-line choices** may be added, if necessary, to the first-line choice. (Refer to the medication formulary for more details). Increased monitoring is required because of sedative effect may precipitate respiratory depression in addition with opioids.
 - Dimenhydrinate 0.5 mg/kg IV q6h PRN, to a max of 50 mg/dose
 - Metoclopramide 0.15 mg/kg IV q4h PRN, to a max of 10 mg/dose
 - Dexamethasone 0.1 mg/kg IV q6h PRN, to a max of 8 mg/dose
 - Droperidol 0.01 mg/kg IV q4h PRN, to a max of 1 mg/dose
- **Pruritus:** **The least sedating agent should always be preferred.**

- **The first-line choice** for opioid-induced pruritus is a naloxone infusion. (Refer to the Medication Administration Protocol for Naloxone for more details.)
 - Using the standard APS solution of Naloxone 10 mcg/mL, give an IV loading dose of 0.1 mL/kg (= 1 mcg/kg) via a volumetric pump. Then start an IV infusion at 0.1 mL/kg/hr (= 1 mcg/kg/hr). This may be increased to 2 mL/kg/hr (=2 mcg/kg/hr) PRN to control pruritus.
- **Second-line choices** may be added, if necessary, to the first-line choice. Refer to the medication formulary for more details). Increased monitoring is required because of sedative effect may precipitate respiratory depression in addition with opioids.
 - Diphenhydramine 0.5-1 mg/kg IV q4h PRN, to a max of 50 mg/dose
 - Nalbuphine 0.05 mg/kg IV q3-6h PRN, to a max of 10 mg/dose. **Warning:** administration to patients who have been receiving high doses of opioids for prolonged periods of time may precipitate acute opioid withdrawal
- **Urinary retention**
 - Any patient receiving regular opioids must be assessed for bladder distension and abdominal pain q4h.
 - If a patient does not void for 8 hours or has symptoms of urinary retention:
 - Using a naloxone solution of 40 mcg/mL (dilute 0.4 mg = 1 mL of naloxone with 9 mL of normal saline for a total of 10 mL), give IV boluses of 2 mcg/kg (0.05mL/kg) q10 minutes x 1-5 PRN until patient voids. If successful in inducing urination, this can be repeated q6-8h PRN. (Refer to the Medication Administration Protocol for Naloxone for more details.)
 - If the patient is still unable to void after the fifth injection, consider inserting a urinary catheter.
- **Constipation:**
 - Eg: Docusate sodium 5 mg/kg/day for a maximum of 400mg/day in one to four divided doses, must be started for all patients who are receiving regular opioids unless contraindicated. Contraindications to docusate sodium include hypersensitivity to docusate or any component, concomitant use of mineral oil, intestinal obstruction, acute abdominal pain, nausea, and vomiting.
 - If the patient has been taking other prescribed medication to treat constipation, this should be continued while the patient is admitted.

f. Discharge to home from ED

▪ **Criteria for Discharge home from the ED**

In order to be considered for discharge home from the Emergency Department from a pain management perspective, patients must meet all of the following criteria:

- Must be able to tolerate fluids
- Must not be requiring IV opioids to maintain an acceptable level of pain control
- Must not have any other contraindications for discharge

ON THE MEDICAL UNIT

- Most frequently admitted patients for sickle cell vaso-occlusive crisis should have a summary sheet of last admission and home pain treatment available in the sickle cell patient binder located in the nurses office on 6C2.
- Assessment of pain, maintenance of analgesia, and treatment of side effects as initiated in the ED (as detailed above) should be continued on the medical unit unless there is a significant change in the patient's pain assessment and/or sedation level.
- May advise (daytime) the Acute Pain Service once the patient is admitted under the medical service. If the medical service feels that a formal consult is more appropriate, this should be done as soon as possible in order for the APS to initiate appropriate treatment in a timely fashion.
- Refer to the **MUHC Pediatric Opioid Therapy Guidelines for monitoring, sedation, and pain scales.**

FOLLOW-UP

Children who are being discharged from hospital after a vaso-occlusive crisis should be seen in hematology/oncology clinic for follow-up within four to six weeks.

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4. APPROVAL PROCESS

Departmental mandate

Department	Print name	Signature	Date
Pediatric Emergency	Dr. H. Eisman	On original	2011/07/19
Medicine	Dr. G. Dougherty	On original	2011/07/05
Nursing	Barbara Izzard	On original	2011/05/17

Institutional and professional approval

Committees	Date
<input checked="" type="checkbox"/> Pharmacy and Therapeutics Paediatrics (if applicable)	2011-04-11
<input type="checkbox"/> Adult Pharmacy and Therapeutics (if applicable)	n/a
<input type="checkbox"/> MUHC Adult Site Medication Administration Policy (MASMAP) (if applicable)	n/a
<input checked="" type="checkbox"/> MUHC Pediatric Medication Administration Policy (PMAP) (if applicable)	2011-04-17
x Nursing Clinical Practice Review Committee (if applicable)	2011-05-09
<input type="checkbox"/> Multidisciplinary Clinical Practice Review Committee (if applicable)	n/a
<input type="checkbox"/> Nursing Executive Committee and Council of Nurses (NEC and CN) (if applicable)	n/a
<input type="checkbox"/> Multidisciplinary Council (if applicable)	n/a
<input type="checkbox"/> MUHC Central Executive Committee of Council of Physicians Dentists and Pharmacists Committee (ECPDP) (Obligatory if attached to a collective order) — Final approval Signature of Chairperson: _____	n/a

5. REVIEW DATE

To be updated in 2016 or sooner if presence of new evidence or need for practice change.

6. REFERENCES

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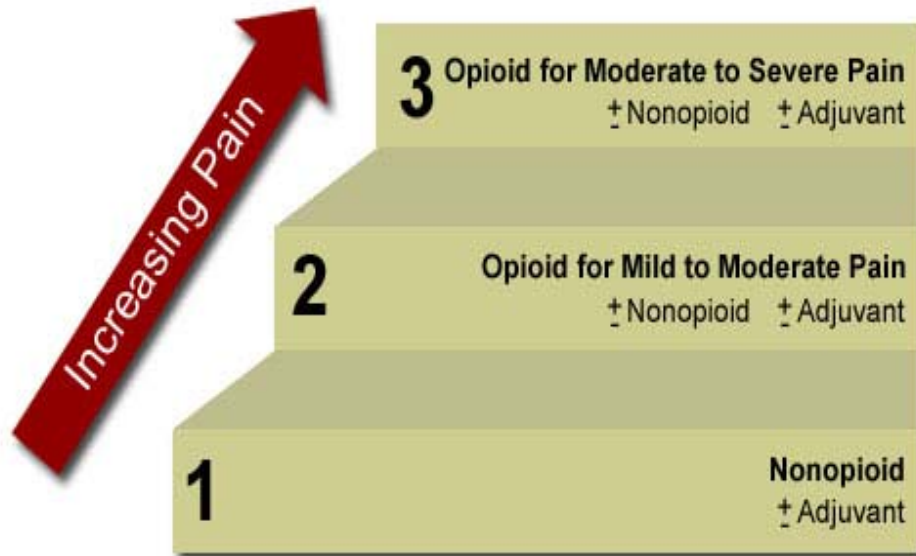
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Appendix I

The WHO Analgesic Ladder



World Health Organization. 1996. Cancer Pain Relief, 2nd Ed.

Appendix II

****This is not a medical order****

Emergency Department Rapid Morphine Titration Algorithm for Sickle Cell Patients > 6 months of age

Physician Guide intended for use in CTAS category 1 and 2 patients

Start acetaminophen 15 mg/kg PO/PR (if not taken in past 4 hrs), repeat Q6H; max single dose = 1 g
AND

Start ketorolac 0.5 mg/kg IVx1 (if no contraindications and no NSAID in past 6 hrs), then 0.3-0.5 mg/kg Q6H; max single dose = 30 mg

