

**INPATIENT MANAGEMENT OF VASO-OCCLUSIVE PAIN  
IN THE CHILD WITH SICKLE CELL DISEASE**

CONSULTS: Hematology

MONITORING:

1. Vital signs with BP q 4 hr. Consider CR monitor.
2. Pain intensity rating at least q 4 hr.
3. Continuous or frequent pulse ox if receiving parenteral opioids or if any respiratory symptoms present.
4. Record I+0. Consider daily weight.

DIAGNOSTICS (if not previously obtained):

1. CBC, diff, platelet count, and retic count initially and daily until improving. (Compare with patient's baseline data.)
2. CXR if cough, thoracic pain, hypoxemia or any respiratory symptoms present or develop after admission. Patients with severe vaso-occlusive pain are at increased risk for acute chest syndrome (see Acute Chest Syndrome Guidelines).
3. If febrile, blood culture and other cultures (e.g. urine, CSF) and urinalysis as indicated.
4. Consider renal (BUN, creat) and liver (fractionated bili, ALT) function tests for very severe pain or any evidence of encephalopathy (R/O acute multi-organ failure syndrome).
5. Consider abdominal ultrasound, liver function tests, and/or amylase and lipase for RUQ, epigastric or severe abdominal pain or marked jaundice (R/O cholelithiasis, cholecystitis, pancreatitis).
6. Type and screen if Hgb is < 6 gm/dl or 15% or more below baseline and/or if evidence of acute chest syndrome (see Acute Chest Syndrome Guidelines) or cardiovascular compromise present. Request leukocyte-depleted and, if available, C, E, Kell-compatible (requires minor antigen phenotype) and sickle-negative RBC. In absence of alloantibodies, urgent transfusion should not be delayed by search for minor antigen matched units.

FLUIDS, GENERAL CARE:

1. IV (D<sub>5</sub> ¼ NS) + P.O. @ 1-1½ x maintenance. Increased fluids may be needed if patient is dehydrated and/or insensible losses are increased (e.g. persistent fever). Avoid excessive fluids, which may precipitate or exacerbate acute chest syndrome.
2. Incentive spirometry - 10 breaths q 2 hr when awake. Consider soap bubbles or pinwheels for younger children.
3. Encourage ambulation and activity.
4. Social work, psychology, child life, and/or chaplain consultation may be helpful.

MEDICATION/TREATMENT: Base choice, dose, and schedule (bolus ATC or PCA) of analgesics in part on severity of pain, analgesics already used, prior experience of patient with efficacy and side effects, and patient preference. In most cases, prn analgesic orders are not appropriate. Never use a placebo. Refer to individual patient care plan if available.

1. Opioid
  - Morphine sulfate 0.05 - 0.15 mg/kg/dose slow infusion IV q 2-3 hr or 0.05 - 0.1 mg/kg/hr continuous infusion or via PCA. (For PCA give 1/2-2/3 of total maximum dose by continuous infusion, with 1/3-1/2 via PCA boluses.) Total morphine dose, continuous infusion plus boluses, above 0.1 mg/kg/hr may be required, especially for opioid-tolerant patients, but should be used with caution.
  - Nalbuphine (Nubain) 0.3 mg/kg IV q 3 hr, 0.2 mg/kg IV q 2 hr, or 0.1 mg/kg/hr continuous infusion. Do not use nalbuphine for patients receiving chronic opioids (e.g. MS Contin, Oxycontin, or fentanyl patch).
  - Other opioids such as hydromorphone (Dilaudid) 0.015-0.02 mg/kg IV q 3-4 hr or fentanyl by continuous infusion or PCA may be appropriate in selected cases. Repeated doses of meperidine (Demerol) should be avoided because of the risk of seizures.
2. NSAID
  - Ketorolac (Toradol) 0.5 mg/kg (30 mg maximum dose) IV q 6 hr or ibuprofen 10 mg/kg po q 6-8 hr if no contraindication present (i.e. gastritis, ulcer, dehydration, coagulopathy, or renal impairment). Limit Ketorolac and more frequent dosing of ibuprofen to 5 days per month maximum duration.
3. O<sub>2</sub> by nasal cannula or face mask as needed to keep pulse ox ≥ 92% or ≥ patient's baseline value, if >92%. The etiology of a new or increasing supplemental O<sub>2</sub> requirement should be investigated. Avoid excessive or unnecessary O<sub>2</sub>, which may suppress the reticulocyte count and exacerbate anemia.
4. If febrile, ceftriaxone 50-75 mg/kg IV q 24 hr (2 gm max single dose) or cefotaxime 50 mg/kg IV q 8 h (2 gm max single dose). Substitute meropenem 20 mg/kg IV q 8 hr (1 gm max single dose) for known or suspected cephalosporin allergy.
5. Add vancomycin 15-20 mg/kg IV q 8 hr (1 gm max single dose) and use higher dose of ceftriaxone (50 mg/kg IV q 12 hr, 2 gm max single dose), cefotaxime (100 mg/kg q 8 hr, 2 gm max single dose), or meropenem (40 mg/kg IV q 8 hr, 2 gm max single dose) for severe febrile illness (e.g., hypotension or poor perfusion) or proven or suspected CNS infection. Draw peak and trough vancomycin levels after 3<sup>rd</sup> or 4<sup>th</sup> dose if vancomycin to be continued > 48 hr.

6. If applicable, continue prophylactic penicillin (should be discontinued while patient is receiving broad-spectrum antibiotics).
7. Offer heating pads, whirlpools, distraction techniques, or other comfort measures previously used by patient. Avoid ice or cold packs.
8. Consider docusate and/or laxative for opioid-induced constipation.
9. Consider diphenhydramine (0.5 mg/kg po q6h, 50 mg/dose max), hydroxyzine (0.5 mg/kg po q6h, 50 mg/dose max), or low-dose nalbuphine (10-20 mcg/kg IV q6h) prn pruritis. Offer menthylated lotion prn itching.
10. Consider promethazine (0.25-0.5 mg/kg po q6h, 25 mg/dose max) or ranitidine (2 mg/kg po q12h, 150 mg/dose max) prn nausea.
11. Consider physical therapy consult for patients with chronic pain or for those hospitalized with acute pain more than 3-5 days.
12. Transfusion is not indicated for uncomplicated episodes of pain. Consider transfusion with RBC if Hgb is <6 gm/dl or 20% or more below baseline, especially with reticulocytopenia, and patient shows any signs of cardiovascular compromise. Request leukocyte-depleted and, if available, C, E, Kell-compatible (requires minor antigen phenotype), and sickle-negative RBC. In absence of alloantibodies, urgent transfusion should not be delayed by search for minor antigen matched units.
13. See other Clinical Guidelines for acute chest syndrome, acute splenic sequestration, aplastic crisis, stroke, priapism, if present.
14. Reassess pain control on a regular basis (at least twice daily and after any change in analgesics) by using age-appropriate pain scale and by discussing efficacy and side effects with patient/family. Analgesics may be weaned as tolerated by decreasing dose, not by prolonging interval between doses. Discuss analgesic changes with patient/family.

DISCHARGE CRITERIA:

1. Adequate pain relief on oral analgesics.
2. Taking adequate oral fluids and be able to take po medications (e.g. prophylactic penicillin) if applicable.
3. Afebrile  $\geq$  24 hr with negative cultures for  $\geq$  24-48 hr if applicable.
4. Resolution of any pulmonary symptoms or documentation of adequate oxygenation on room air.
5. Stable hemoglobin/hematocrit
6. Follow-up arranged.

*These guidelines do not indicate an exclusive course of treatment or serve as a standard of care. Variations based on a physician's best medical judgement may be appropriate in individual cases.*