INPATIENT MANAGEMENT OF PAIN EPISODE IN SICKLE CELL DISEASE (Jan 2009)

ADMIT: Under Ped Hematology - (352) 392-5633 to speak with hematologist (8a-5p).

MONITORING: **If respiratory symptoms** are present ***see*** **Acute chest syndrome (ACS) guidelines**

1. **Vital signs, BP q 4hr**. Consider CR monitor.
2. **Pain intensity rating at least q 4 hr**.
3. **Continuous pulse ox,** if getting IV opioids.
4. **Strict I+0**. Consider daily weight.

DIAGNOSTICS (if not previously obtained):

1. **CBC, diff, platelets & retic** count initially & daily until improved. (Compare with pt's baseline)
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 function tests (fractionated bili, ALT) 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/upper abdominal pain or marked jaundice (R/O cholelithiasis, cholecystitis, pancreatitis, splenic sequestration – see sickle cell handbook).
6. Type and screen if Hgb is < 6 gm/dl or 25% or more below baseline and/or ACS (see ACS Guidelines) especially if symptomatic or with new/worse splenomegaly (see blood transfusion and splenic sequestration guidelines) or with disproportionate reticulocytopenia. 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_5½ 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. Use soap bubbles or pinwheels for younger kids.
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. 0_2 by nasal cannula or face mask as needed to keep pulse ox $\geq$ 92% or $\geq$ patient's baseline value, if >92%. The etiology of a new or increasing supplemental 0_2 requirement should be investigated. Avoid excessive or unnecessary 0_2, which may suppress the reticulocyte count and exacerbate anemia.
4. If febrile, ceftriaxone 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). (Refer to fever guidelines).
5. If applicable, continue prophylactic penicillin (should be discontinued while patient is receiving broad-spectrum antibiotics).
6. Offer heating pads, whirlpools, distraction techniques, or other **comfort measures** previously used by patient. Avoid ice or cold packs.
7. Consider docusate and/or **laxative** for opioid-induced constipation.
8. **Pruritus**: 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). Offer menthylated lotion prn.
9. **Nausea**: 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.
10. Consider **physical therapy consult** for patients with chronic pain or for those hospitalized with acute pain more than 3-5 days.
11. 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.
12. For **other co-morbidities** see Clinical Guidelines for acute chest syndrome, acute splenic sequestration, aplastic crisis, stroke, priapism, if present.
13. **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.