MANAGEMENT OF ACUTE CHEST SYNDROME WITH SICKLE CELL DISEASE (Jan 2009)

DEFINITION: An acute illness with fever, respiratory symptoms and new pulmonary infiltrate on CXR.

CONSULTS: Admit under Pediatric Hematology, consider Pulmonology/PICU

MONITORING:

1. *Hospitalize*.
2. *Vital signs with BP q 2-4 hr*.
3. *Pain intensity rating at least q 4 hr*.
4. *Continuous pulse oximetry*.
5. *Strict I+O, daily weight*.
6. Consider PICU if severe/rapid deterioration.

DIAGNOSTICS:

1. *CBC, diff, platelet count, and retic* count initially and daily until improving. Compare with patient's baseline values.
2. *CXR* initially, repeat for significant changes or clinical deterioration.
3. Consider:
   - *Type and crossmatch* for severe illness or if Hb <6 gm/dl or >15% below baseline especially if symptomatic or with new splenomegaly (see blood transfusion and splenic sequestration guidelines) or with disproportionate reticulocytopenia. Request leukocyte-depleted & C, E, Kell-compatible, sickle-negative RBCs. In absence of alloantibodies, emergent transfusion should not be delayed by search for minor-antigen matched units.
   - *Blood cultures* if febrile or history of recent fever (<24 hrs) and q 24 hr, if fever > 38.3^0C.
   - Consider *blood gas* or arterial line, PICU for CPAP or BIPAP for severe illness.
   - *Renal* (BUN, creat) and *liver function tests* (fractionated bili, ALT) for severe illness or if diffuse encephalopathy present (R/O acute multiorgan failure syndrome).

FLUIDS, NUTRITION, GENERAL CARE:

1. Maintain "euvolemia." *IV (D_5 ½NS) + P.O. @ 1x maintenance*. More fluid is appropriate only if patient is dehydrated or if insensible losses are increased (e.g. persistent fever).
2. *Incentive spirometry* - 10 breaths q 2 h when awake. Consider soap bubbles or pinwheels for younger patients. Consider chest PT for significant pneumonia, or inability to perform above.
3. Encourage *ambulation*, activity.

MEDICATIONS/TREATMENTS:

1. *Oxygen* to keep pulse ox ≥ 92% or ≥ baseline value, avoid using oxygen for 'comfort'.
2. *Albuterol* nebs q 4-6 hr, especially if patient has h/o reactive airway disease or wheezing. Consider *CPT* or vibratory vest. Continue outpt asthma meds (e.g., Advair) if applicable.
3. Consider *PRBCs or PICU* for positive pressure ventilation (CPAP or mask BIPAP) for patients with poor or increasing respiratory effort/reduced ventilation/increased oxygen requirement.
4. Consider red cell transfusion:
   - *Simple transfusion* for moderately severe illness, especially if Hb >1 gm/dl below baseline (do not transfuse acutely to Hb >10 gm/dl, Hct >30%).
   - *Partial exchange* transfusion to Hb 10 gm/dl and Hb S or Hb S+C (patient's RBC) ≤ 30% for severe or rapidly progressive disease (may require transfer to ICU and transfusion medicine consult for erythrocytapheresis). Remove femoral or central venous catheters as soon as possible after exchange transfusion to reduce risk of thrombosis.
5. *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). Substitute Clindamycin 10 mg/kg IV q 6 hr (max dose 4800 mg per day) or quinolones or meropenem 20 mg/kg IV q 8 hr (1 gm max single dose) for patient with known or suspected *cephalosporin allergy*. Increase ceftriaxone to 100 mg/kg/day if resistant pneumococcus is suspected. Prophylactic penicillin should be discontinued while patient is receiving broad-spectrum antibiotics.
6. *Azithromycin* 10 mg/kg po (500 mg max single dose) x 5 days or other macrolide antibiotic.

7. Strongly consider *adding vancomycin* 15-20 mg/kg IV q 8 hr (1 gm max single dose) for severe illness or for suspected *S. aureus* infection. Draw vancomycin trough levels after 3rd or 4th dose if vancomycin is to be continued > 48 hr.
8. Opioid
   - *Morphine* sulfate 0.05 - 0.15 mg/kg/dose IV q 2-3 hr or 0.03 - 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. Recheck ½ hr after dose for pain control/excess sedation.
   - Nalbuphine (Nubain) 0.1 mg/kg IV q 2-3 hr, or 0.03 - 0.1 mg/kg/hr continuous infusion. Do not use Nubain for patients receiving chronic opioids (e.g. MS Contin, Oxycontin).
   - *Other opioids* such as hydromorphone (Dilaudid) 0.015-0.02 mg/kg IV q 3-4 hr or fentanyl may be appropriate in selected cases. Repeated doses of meperidine (Demerol) should be avoided because of the risk of seizures and nephrotoxicity.
9. *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 is present (i.e. gastritis, ulcer, dehydration, coagulopathy, or renal impairment). Limit Ketorolac and Ibuprofen to 5 days per month.
10. *Incentive spirometry* 10 times q 2 hrs when awake.
10. Avoid furosemide (<0.5 mg/kg IV if fluid overload). *Consider fluid restriction* instead.
11. Acetaminophen 10-15 mg/kg po q 4 hr or prn T >38.3^{o}C (75 mg/kg/day or 4 gm/day max).
12. Consider ducosate and/or laxative for opioid-induced *constipation*.
13. For *pruritis*, consider diphenhydramine (0.5 mg/kg po q6h, 50 mg/dose max, excessive sedation is likely with morphine), hydroxyzine (0.5 mg/kg po q6h, 50 mg/dose max), or low-dose nalbuphine (10-20 mcg/kg IV q6h). Offer menthylated lotion.
14. For *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) for gastritis.
15. Offer heating pads or *other comfort measures* previously used by patient. Avoid ice/cold packs.
16. *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.
17. Watch for *other co-morbidities* - acute cholecystitis, gastritis, acute splenic sequestration, aplastic crisis, stroke, priapism – please see sickle cell handbook for specific guidelines.

DISCHARGE CRITERIA:

1. Improved pulmonary symptoms and documentation of adequate oxygenation on room air.
2. Afebrile ≥ 24 hr. and negative cultures for ≥ 24-48 hr if applicable.
3. Stable hemoglobin/hematocrit.
4. Taking adequate oral fluids and able to take po medications if applicable.
5. Adequate pain relief, if needed, with oral analgesics.
6. Follow-up plans coordinated with hematology service. On a case by case basis, consider follow-up pulmonary function testing or chronic transfusions or hydroxyurea.

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.