Inpatient Sickle Cell Disease Acute Chest Syndrome Guideline

**Fever, cough, dyspnea, wheezing, chest pain**

- **Begin Pain Management Immediately**
- Patients should be treated according to their individualized management plan (Refer to ED binder).

**History**
- Obtain and document associated symptoms
- Review any oxygen requirement, obtain baseline Oxygen sats.
- Review history of asthma, current medications, drug allergies
- Review medical history focusing on sickle cell disease, history of acute chest syndrome and pneumonia

**Laboratory Evaluation**
- CBC (including WBC diff and platelet)
- Rapid HgbS level
- Chemistry profile
- Liver function tests
- Type and Screen (sickle cell negative-leukocyte depleted)
- PRBC’s, Rh (C,E,) and Kell antigen-matched blood. (Patients with prior history of allosensitization should receive PRBCS that are phenotypically identical.)
- Consider ABG based on clinical condition (Severe hypoxemia PaO2 < 70 mm Hg indicates potentially life-threatening disease.)

**Transfusion Therapy**
- Hct < 30 %= Red cell transfusion
- Multiple lobe involvement, PaO2 < 70 mm Hg= Perform an exchange transfusion
- Early transfusions are indicated for patients at high risk for complications.

**Monitoring**
- Continuous pulse oximetry
- Vital signs with blood pressure at least every 4 hours
- Consider cardio-respiratory monitor if suspect over-sedation
- Chest x-ray daily
- Accurate Intake and Output

**Management**
- Oxygen to maintain sats ≥ 94%
- Bronchodilators
- Chest X-ray daily and with clinical deterioration
- Incentive spirometry every 2 hours while awake, every 4 hours while sleeping.
- Consider chest physiotherapy
- ABg<70, perform exchange transfusion – respiratory failure likely
- Hydration using oral and IV fluids. Total hydration should not exceed 1-1 ½ times maintenance.
- I/O and weigh daily

**Medications**
- Analgesics for pain control (refer to pain guideline)
- Appropriate antibiotic combination of cephalosporin (Ceftriaxone) and macrolide (Azithromycin) - see formulary for dosing details.
- Consider adding Vancomycin if clinical deterioration
- Continue prophylactic penicillin
- Tylenol 10-15 mg/kg/dose for fever
- Laxatives for narcotic-induced constipation.
- For puritis administer antihistamines such as diphehydramine or hydroxyzine
- Antiemetics for nausea

**Discharge home with previous pain control regime when meets the discharge criteria of:**
- Sats ≥ 94% on room air or baseline O2 requirement
- Afebrile for at least 24 hours
- Adequate oral intake, including medications
- Adequate pain relief (if needed) with oral analgesics

---

**Guideline developed by Lakshmanan Krishnamurti, MD & staff**