Sickle Cell Disease (SCD)
Acute Chest Syndrome Guidelines

Background

Acute chest syndrome (ACS) is the second most common cause for hospitalization and leading cause for death in sickle cell disease. The peak incidence of ACS occurs in children between 2 and 4 years of age with a higher prevalence during the winter months. Risk factors for ACS include: History of asthma, infection from atypical organisms (Chlamydiae and Mycoplasma sp.), hypoventilation from atelectasis secondary to surgery, pain from rib infarction or the excessive administration of opioid therapy.

ACS is defined by the presence of a new chest X-ray infiltrate plus one of any respiratory symptoms (chest pain, temperature >38.5°C, tachypnea, wheezing, coughing or new-onset hypoxemia)

Laboratory Studies, Chest X-ray Inpatient

Daily CBC with differential
Daily Reticulocyte count
Blood culture for ongoing fever
Rapid flu during influenza season (if not already done in the ER)
Chest radiograph – repeat if clinically indicated
- Worsening respiratory symptoms
- New hypoxia

Inpatient Management:

Maintenance IVF
Continuous pulse oximetry
Incentive spirometry q 2 to 4 hours during the day, and q vitals during the night
Blowing into bubbles q 2 to 4 hours in young children unable to perform spirometry
Oxygen as needed for hypoxia to keep O2 saturations >93%
Consider transfusing packed red blood cells 10cc/kg over 2 hours
Albuterol scheduled for presence of wheezing (what about patients without wheezing)
Refer to inpatient pain management guidelines for treatment of pain
Azithromycin for atypical organism coverage
Consult pulmonary
Continue other home asthma medications (if patient has history of asthma)

Outpatient Follow-up

Make follow-up appointment with Pediatric pulmonary (Dr. Josey)
Optimize hydroxyurea therapy
Follow-up as scheduled with CCC