Respiratory Symptoms in Patients with Sickle Cell Disease

Developed by the NC Division of Public Health; the comprehensive sickle cell centers at Carolinas Health Care, Duke University, East Carolina University, University of North Carolina at Chapel Hill, Mission, and Wake Forest University; Endorsed by (SC)².

Respiratory Distress?

Transfer to emergency facility. Give bronchodilator, if wheezing, while awaiting transfer.

If possible, give parental antibiotic coverage for strep pneumonia and gram negative enteric pathogens (e.g., IM Ceftriaxone) prior to transfer if concern for Acute Chest Syndrome*.

*Acute Chest Syndrome – Any new infiltrate with clinical symptoms. *CXR may be negative in the first 24 hours of symptoms. Typically, sudden onset of s/sxs of lower respiratory tract disease (e.g., cough, shortness of breath, retractions, rales, etc.) and a new pulmonary infiltrate on chest radiograph. Hemoglobin concentration often declines sharply below the patient’s baseline value. In the early stages of ACS, the clinical manifestations can be subtle. Children usually have fever and upper or middle lobe involvement. Adults often afebrile and present with multilobe disease. Most common etiology is infection (e.g., viral, bacterial, chlamydia, or Mycoplasma), but may also result from bone marrow fat embolism, intrapulmonary aggreage of sickled cells, atelectasis, or pulmonary edema.