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; and primary care physicians from across North Carolina. Adapted from the 2014 NIH/NIH guidelines for Evidence-Based Management of Sickle Cell Disease.

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*.

Signs/sxs of pneumonia, ACS (e.g. crackles, fevers)?

CXR

Close follow-up. See fever algorithm if temp >100.5°

Any signs and symptoms indicative of pneumonia or Acute Chest Syndrome* (e.g. crackles, fever, and/or positive chest x-ray)?

Consider re-assessment in 24 hours

Stable at 24-hr follow up?

Yes

Chronic Hypoxia. Refer to sickle cell specialist and/or pulmonology

No

Consider re-assessment in 24 hours to assess if stable

Yes

Stable at 24-hr follow up?

No

Pulse ox significantly below baseline or <92% (children), <90% (adults)?

Yes

Bronchodilator treatments

No

Take pulse ox and temperature

Wheezing?

Yes

Wheezing, symptoms, Hypoxia improved?

Close follow-up. See fever algorithm if temp >100.5°

No

No

Close follow-up.

Yes

Patient presents with respiratory symptoms

Respiratory Distress?

Yes

No

No

Signs/sxs of pneumonia, ACS (e.g. crackles, fevers)?

CXR

Close follow-up. See fever algorithm if temp >100.5°

No

No

Yes

Yes

Consider re-assessment in 24 hours

*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 aggregates of sickled cells, atelectasis, or pulmonary edema.