

CLINICAL ALGORITHM FOR MANAGEMENT OF ACUTE CHEST SYNDROME IN PEDIATRIC SICKLE CELL PATIENTS

This pathway should be used for patients > 2 months of age with SS, SC, or S β -thalassemia type sickle cell disease who present with concerns for acute chest syndrome. Please direct questions to Maine Children's Cancer Program (207-396-7565)

Management of Acute Chest Syndrome in Pediatric Sickle Cell Patients

Acute chest syndrome is characterized by a new pulmonary infiltrate detected by chest radiograph **and** one or more of the following: temperature ≥ 38.5 , cough, chest pain, tachypnea, retractions, wheezing and hypoxemia
Evaluation for acute chest should include history, physical and the following diagnostic workup :

- Chest x-ray
- CBC with differential
- Reticulocyte count
- Comprehensive metabolic panel
- Type and screen
- Blood culture if febrile
- Consider RSV, flu testing, respiratory viral culture

Initial management

The primary infectious agents implicated in ACS include: Chlamydia pneumoniae, Mycoplasma pneumonia and Streptococcus pneumonia. Other causes of ACS include bone marrow fat embolism, intrapulmonary aggregates of sickled cells, atelectasis, and pulmonary edema.

- **Call pediatric hematology/oncology for admission**
- Ceftriaxone 50mg/kg IV or IM **AND** Azithromycin 10mg/kg PO or IV on day 1 followed by 5mg/kg days 2-5
- For cephalosporin allergy replace with Clindamycin 40mg/kg/day divided q6-8hrs
- For the severely ill patient consider adding Vancomycin with pharmacy consult
- Prophylactic penicillin should be discontinued while receiving antibiotics
- Oxygen to keep O₂ sat $\geq 95\%$
- Consider bronchodilators if history of asthma or evidence of bronchospasm
- Correct hypovolemia with isotonic fluid (careful to avoid fluid overload)
- Fever management with acetaminophen
- Adequate pain management
- Incentive spirometry or other age appropriate modality (e.g. blowing bubbles)

Simple Blood Transfusion

Goal is to improve oxygenation and prevent progression to respiratory failure

Consider transfusing **5-10cc/kg** of sickle negative, extended antigen-matched (if available) pRBC's for symptomatic patients if:

- Hb concentration > 1.0 g/dL below baseline and Hb < 9 g/dL
- Clinical or radiological progression

* Try not to exceed Hct of 30% or Hb >10g/dL post transfusion

In patients with HbSC disease or HbS β -thalassemia with ACS, decisions about transfusion should be made in consultation with pediatric hematologist

Exchange Transfusion

Goal is to remove sickled cells (Hb S) and replace with Hb A. Consider exchange transfusion if:

- Critically ill
- Multilobe involvement or progressive pulmonary infiltrates
- Decline in Hb despite simple transfusion
- Hb ≥ 9 g/dL
- Increasing respiratory distress despite initial management

*Exchange transfusion requires consult to pediatric hematology, pediatric nephrology and PICU admission



Algorithms are not intended to replace provider's clinical judgment or to establish a single protocol. Some clinical problems may not be adequately addressed in this guideline. As always, clinicians are urged to document management strategies.