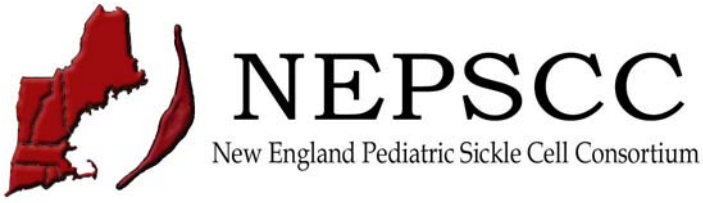
New England Pediatric Sickle Cell Consortium



Management of Acute Chest Syndrome in Sickle Cell Disease

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Disclaimer Statement:

- Hospital clinical pathways are designed to assist clinicians by providing an analytical framework for the diagnosis and treatment of specific medical problems. They may be used for patient education and to assist in planning future care. They are not intended to replace a physician's judgment or to establish a protocol for all patients with a particular condition. The ultimate decision regarding the care of any patient should be made in respect to the individual circumstances presented by the patient.
- Any specific medications and dosing must always be reviewed carefully for each patient in view of any history of drug allergy or adverse reactions.
- This document was based on available research and clinical experience at time of its compilation.
- The following protocol is a regional guideline, and may be adapted by individual institutions as needed.

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Acute Chest Syndrome

Inclusion:

I. Introduction
II. Evaluation and Management
III. Follow-Up and Prevention

References

I. Introduction

Inclusion: all children and adolescents with sickle cell diseases

Definition: a new infiltrate on CXR, excluding atelectasis, plus one or more of the following1

- tachypnea
- fever > 38.5
- chest pain
- cough
- wheezing
- hypoxemia (room air SaO2 3-5% points less than baseline)

- ACS is a leading cause of morbidity and mortality in children and adults with SCD2
- More common in HbSS, and in those with baseline low Hb, % HbF and high WBC3
- Patients with frequent episodes of ACS and VOC have shorter lifespans
- Potential etiologies include infarction, infection and pulmonary fat embolism4
- Infectious agents most often identified are chlamydia, mycoplasma and viruses5
- Most common initial presentation is fever (80%)
- More than ½ of the episodes of ACS are complicated by VOC events
- VOC involving the trunk cause splinting and decreased tidal volume, and are associated with ACS episodes
- ACS is the most common complication of surgery and anesthesia in patients with SCD6
- Studies have shown SCD patients to have hyperreactive airways and more lower airway obstruction
- Patients with asthma are more likely to progress to ACS following VOC7
- In adults, treatment with hydroxyurea has reduced episodes of ACS by 50%8
- Pediatric patients on chronic transfusions therapy have significantly fewer episodes of ACS9
- Use of incentive spirometry has been shown to decrease incidence of ACS in patients hospitalized with VOC10
- Systemic steroids have been shown to hasten recovery from ACS, but are also associated with rehospitalization from rebound VOC11
- Complication of ACS is mucous plugging which can lead to obstruction, the severe form is known as 'plastic bronchitis'12
- Severe episodes of ACS have been associated with neurological damage.13 14

II. Evaluation & Treatment

Patients with acute chest syndrome may present with fever, pain of the chest or abdomen, asthma exacerbation or other respiratory symptoms.

A. Initial Evaluation
- If febrile, perform work-up as per Fever CPG
- All patients must have blood counts, blood culture and a type & screen to blood bank
- Any SCD patient with a new infiltrate on CXR requires hospitalization for at least 1 night

B. Respiratory
1. Oxygen: if SaO2 is < 3-5% points below baseline
2. Treatments
- Incentive spirometry 10x/h while awake, and may wake to use overnight if hypoxic
- Encourage getting out of bed and ambulation
- Chest physiotherapy if clinically indicated, and tolerated
- BiPap overnight may be helpful, especially if splinting (where available)

3. ABGs
- for significant respiratory distress
- to determine need for exchange transfusion
- consider to verify oxygen requirement, especially if SaO2 is inconsistent with clinical picture

C. Fluid-Volume
- Correct any fluid deficit then IV+PO= 1x maintenance; do not exceed 1.5x maintenance, even if concurrent VOC
- Do not bolus unless clinically dehydrated
- Caution not to over-hydrate or rapidly hydrate as fluid overload may contribute to respiratory distress
- Consider diuretics if fluid overloaded or fluid sensitive

D. Medications
1. Antibiotics
- Broad spectrum parenteral (e.g. Ceftriaxone, Cefuroxime)
- Plus oral macrolide (e.g. Azithromycin for 5 days)
- May add Vancomycin for severe disease not responsive to treatment

2. Bronchodilators:
- For active wheezing
- Any patient with history of reactive airway disease (RAD)
- As trial for any patient with ACS, continue if clinical response
- Caution that albuterol may cause arrhythmias when used with drugs that prolong the QT interval (erythromycin)

3. Steroids:
- For symptomatic RAD or severe respiratory compromise
- Steroids can cause rebound VOC and may need to be tapered
- Dexamethasone (Decardon) at 0.3 mg/kg/dose q 12 hours x 2 days then taper
- May treat those with RAD with prednisone at 1-2 mg/kg/day x 5 days then taper

4. Pain
- Optimize pain control to decrease chest splinting and prevent atelectasis
- Avoid hypoventilation from oversedation
- Use NSAIDs alone or in conjunction with opioids
- Consider trial of stimulants if excessive sedation from opioids

E. Transfusion Considerations (see Transfusion CPG)

1. Straight/simple

- Goal is to improve oxygenation and prevent progression to respiratory failure
- For patients with accentuated anemia (>10-20% drop from baseline)
- For patients who are symptomatic, but not in impending respiratory failure
- For patients with clinical or radiological progression
- For patients with in which exchange will be delayed, simple transfusion may temporize.
- Try not to exceed Hct of 30% or Hb >10g/dL post transfusion

2. Exchange/erythropheresis

- Goal is to remove sickled cells (Hb S) and replace with Hb A
- For impending or actual respiratory failure
- For marked clinical deterioration with progressive radilogical findings, especially after a simple transfusion
- For patients with hb ≥10gm/dl or hct ≥30% to avoid the increased viscosity of a simple transfusion

F. Options for Respiratory Failure15

1. Exchange transfusion
2. Respiratory support
   - Conventional mechanical ventilation
   - High-frequency oscillatory ventilation
     - For ventilated patients repeated daily bronchoscopy with lavage and aggressive suctioning of bronchial casts
   - Nitric Oxide (NO) via mask or endotrachial tube
   - Extra Corporeal Membrane Oxygenation (ECMO) has been reported
3. Medication
   - Ensure broad antibiotic coverage
   - Systemic corticosteroids (methylprednisone, dexamethasone)
   - Bronchodilators
   - Inhaled mucolytics
4. Neurological: Monitor for signs of neurological complications. Stroke is associated with ACS

G. Labs and Chest X-rays:

- Daily CXR if lack of clinical improvement; more often if clinically worsening
- Daily CBC with differential and reticulocyte count
- Consider viral studies in winter/spring months
- Consider mycoplasma titer or cold agglutinins if clinically appropriate

H. Discharge Criteria

- Improved respiratory status with stable blood counts
- CXR not worsening
- Family able to care for child at home

I. Discharge Medications

- Complete course of oral antibiotics
- Treatment for RAD, if started in the hospital
- If steroids are used, they should be tapered

J. Follow-Up

- Appointment to be given before discharge
- Should be seen within 3-5 days if treated with steroids
- Repeat CXR in 6 weeks
- Consider f/u PFTs in 3 months for patients with severe episode

- If transfused send blood bank specimen in 6-8 weeks for allosensitization

III. Prevention

A. Pulmonary

- Follow-up in Pulmonary or Allergy/Asthma clinic for those with comorbid asthma.
- For hospitalized patients with VOC encourage ambulation and frequent use of incentive spirometry.
- Families must understand importance of seeking prompt evaluation for respiratory symptom.

B. Infectious Diseases

- Ensure vaccinations given for pneumococcal disease
- Yearly influenza vaccine after age 6 months
- Encourage influenza vaccination for household contacts

C. Hematology

Patients with non-modifiable risk factors (severe anemia, high WBC, recurrent or single severe ACS) should be considered for:

1. Hydroxyurea (see Hydroxyurea CPG for implementation)
2. Chronic Transfusion (see Transfusion CPG for implementation)
    - Must consider risk/benefit of transfusion therapy including iron overload, alloimmunization, autoantibodies
    - Short term: up to 6 months with consideration of concommittant transition to Hydroxyurea
    - Long term: more than 6 months
3. Stem Cell Transplantation. May be appropriate if chronic lung disease or recurrent, severe ACS episodes

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