

# Management of Acute Chest Syndrome in Children with Sickle Cell Disease

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Any patient with sickle cell disease who has fever, tachypnea, hypoxia, retraction or nasal flaring  
(If patient is febrile, the timing of blood culture and first dose of antibiotic administration should be according to fever and sickle cell disease guideline)

Radiographic changes can lag behind clinical findings

Normal physical exam in 25 % of cases

Positive physical findings or positive findings on CXR

Negative physical findings and negative CXR

CBC, Retic, CMP, B/C  
Treat for ACS

CBC, retic, Blood Culture  
Continue Observation  
Frequent examination

Admission to inpatient floor

Vital signs q 4 hours

Continuous pulse ox

Oxygen therapy

Strict I&O

Daily weight

Incentive spirometry 10 breaths q 2 hours while awake

Encourage ambulation

IVF's- 1- 1 ½ maintenance

Pain management

Start Antibiotics

Hold prophylactic antibiotic

Bronchodilator

Corticosteroid

Oseltamivir if indicated

Blood transfusion (simple or exchange transfusion)

### Transfer to PICU

↓ O2 saturations

↑ O2 requirements

increasing shadowing on chest x-ray.

Falling HB

BiPAP

Assisted ventilation

Nitrate oxide

Exchange transfusion

### Discharge from hospital

- Temperature <38 °C for 24 hours without IV antibiotics.

- Normal, stable oxygen saturation measurements, as assessed by pulse oximetry

- If continuing oral antibiotics, instructions to resume prophylactic Penicillin V when finished

- Reinforce to Start or continue hydroxyurea

**Population:** this guideline is aimed at all children in Cardinal Glennon Children's Medical Center with sickle cell disease and respiratory symptoms where the acute chest syndrome may be the cause.

**Definition:** Acute chest syndrome (ACS) is defined as the presence of a new pulmonary infiltrate, irrespective of the etiology on a chest x-ray, in a child with sickle cell disease (SCD).

**Background:** ACS is the second most common cause of hospital admission in children with SCD, and the commonest cause of death. ACS is multifactorial in origin, with contributions from infection, vaso-occlusion, fat embolism, and disturbed nitric oxide metabolism. The prevalence of both viruses and atypical bacteria as common causes of infection suggest that the clinician must carefully consider the antimicrobial agents prescribed, which should provide coverage against atypical bacteria.

### **History and Physical Examination:**

Some of the following symptoms and signs are typically present:

#### **Symptoms:**

- Chest pain, which may be absent particularly in younger children
- Cough, which may be productive
- Breathlessness
- Wheezing
- Fever/rigors

#### **Signs:**

- Fever
- Tachypnea
- Wheeze, crackles
- Bronchial breathing
- Cyanosis
- Tachycardia
- Hypoxia
- Retractions
- Nasal flaring
- Abnormal breath sounds- rhonchi, decreased, pleural rub, crackles

\*Rationale: 25% of patients with Acute Chest Syndrome (ACS) have a normal physical examination, and 60% are not suspected on clinical basis.

**Diagnostic evaluation:**

- CBC with differential, Reticulocyte count
- CXR
- Consider Type and Screen
- Basic biochemistry (creatinine and liver function tests)
- Blood cultures
- VBG /ABG measurement depends to physician discretion
- Respiratory viral panel (PCR- viral resp screen)
- Pulse oximetry in room air
- CBC and retic should be checked daily until patient improves

**Monitoring and treatment:**

- The hematology team should be informed.
- All patients with ACS should be admitted to hospital.
- The critical care team should also be made aware of the patient, even in mild cases of ACS, because clinical deterioration often occurs rapidly and unexpectedly.
- Monitor vital signs every 4 hours until patient improves.

**Oxygen:** All patients should be given oxygen to maintain their oxygen saturations above 95%.

**Physiotherapy:** Incentive spirometry 10 breaths q 2 hours while awake may help prevent children with acute pain developing acute chest syndrome.

**Intravenous Fluids:** In general, all patients with ACS should receive intravenous fluids at the maintenance rates, which may need to be modified according to fluid loss and fever. If patients are considered to be dehydrated and require higher rates of iv fluid replacement this must be reviewed within 12 hours and reduced once patients are adequately hydrated, to reduce the risk of fluid overload which can complicate ACS and lead to clinical deterioration.

**Antibiotics:** The most common bacterial organism in children is *Mycoplasma pneumoniae* and the commonest virus identified is the respiratory syncytial virus (RSV). *Staphylococcus aureus*, *Streptococcus pneumoniae*, *Haemophilus influenzae* and respiratory viruses other than RSV are also seen.

Prophylactic antibiotic should be held while the patient is on antibiotics for treatment of ACS.

If patient is febrile, the recommended or optimal time to initial antibiotic administration is <60 minutes. Administer once cultures are obtained, without waiting for lab results.

**Ceftriaxone:** 50mg/kg IV every 24 hours. (Maximum dose =2 gram)

**Azithromycin:** 10mg/kg PO for the first day and then 5/mg/kg /day for the next 4 days. (Maximum dose of 500 mg)

**Special considerations:**

**Vancomycin:** 10-15mg/kg IV every 6 hours for severe illness, pleural effusion

**Antivirals:** if influenza A is positive: **Oseltamivir** (oral for 5 days), <15kg = 30mg PO BID,

16-23kg= 45mg PO BID, 24-40kg= 60mg PO BID, >41kg=75mg PO BID

**Bronchodilators-**: Nebulized albuterol should be used in all patients initially and continue if there is wheezing or history of reactive airway.

**Corticosteroid:** Can be used in patients with history of asthma, lack of improvement after 4 days of treatment or deteriorating patients.

**Analgesia:** Pain should be treated according to CGCH guidelines.

### **Deteriorating Patients:**

Patients with ACS can deteriorate rapidly and require close monitoring.

PICU should be alerted that a deteriorating ACS patient is on the floor. Up to 10% may need ventilator support.

Deterioration is suggested by:

- Decreasing level of consciousness
- Decreasing oxygen saturations (measured by pulse oximetry) in air
- Increasing oxygen requirements to maintain 100% oxygen saturations, or failure of oxygen to correct saturations.
- Increasing tachypnea
- Increasing pain
- Fluid balance - assess for possibility of fluid overload
- Opiate analgesia – assess for possibility of opioid toxicity
- Increasing shadowing on chest x-ray
- Falling hemoglobin, increasing white cell count

### **Blood transfusion**

- Simple **blood transfusion:** Consider simple blood transfusion (10 mL/kg RBCs):
  - If Hb concentration is >1.0 g/dL below baseline, especially if baseline is less than 9 g/dl.
  - In hypoxic patients with Hb less than 9g/dl.
- Exchange **transfusion:** This is indicated in rapidly deteriorating patients, particularly with extensive chest x-ray shadowing and low oxygen saturations and/or decline in hemoglobin concentration (despite simple transfusion) or in those with higher hemoglobin concentration (>9 g/l).

### **Discharge from Hospital:**

- Temperature < 100.4 F C for 24 hours without IV antibiotics

- Normal, stable oxygen saturation measurements, as assessed by pulse oximetry, i.e. equivalent to pre-morbid reading
- Normal respiratory rate
- If continuing oral antibiotics, instructions to resume prophylactic Penicillin V when finished
- An outpatient appointment arranged with the sickle cell and pulmonary clinics
- Advice about outstanding immunizations including Pneumovax, Meningococcal, and Annual Influenza vaccine
- Reinforce about starting or continuing hydroxyurea.

## **References:**

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