1.0 Introduction

Acute chest syndrome (ACS) is defined as a new infiltrate on chest x-ray associated with new respiratory symptoms and is responsible for up to 25% of all deaths in children with sickle cell disease, and is the second most common cause for hospitalization in these children. The etiology of ACS is variable and may include both infectious and non-infectious causes; infections are more common in younger children. (Organisms include but are not limited to those listed below.)

<table>
<thead>
<tr>
<th>Infectious Causes</th>
<th>Non-infectious Causes of ACS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bacteria</td>
<td>• Pulmonary infarction <em>(in situ</em> sickling)</td>
</tr>
<tr>
<td>• Pneumococcus</td>
<td>• Hypoventilation secondary to rib/ternal infarction or narcotic administration</td>
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<tr>
<td>• Gram-negative bacteria</td>
<td>• Fat embolism</td>
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<tr>
<td>• Chlamydia pneumoniae</td>
<td>• Pulmonary edema secondary to fluid overload</td>
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<tr>
<td>• Mycoplasma pneumoniae</td>
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<tr>
<td>Viruses</td>
<td></td>
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<tr>
<td>• Respiratory syncytial virus</td>
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<tr>
<td>• Para-influenza</td>
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<tr>
<td>• Influenza</td>
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</tbody>
</table>

In patients with sickle cell disease, ACS occurs most frequently in patients with haemoglobin genotype SS (12.8 events/100 patient-years); less so in those with HbSβ0 thalassemia (9.4 events/100 patient-years) or HbSC (5.2 events/100 patient-years); and least often in those with HbSβ+ thalassemia (3.9 events/100 patient-years) (Castro et al. 1994). Within each Hb type, the incidence is strongly but inversely related to age, being highest in children 2–4 years old (25.3 events/100 patient-years) and decreasing to its lowest value in adults.

2.0 Clinical/Laboratory Features

Frequency of presenting symptoms in ACS appears to be age-specific. In young children (2-4 years old), fever and cough are typical; pain is rare; and upper lobe disease is more common. Adults tend to present with shortness of breath, chills, severe pain, and no fever; multi-lobe and lower lobe disease are more frequent. Seasonal variation is seen, with more cases reported in the winter.

Tenderness may be present over the ribs or sternum. Chest x-rays of patients with ACS may show infiltrates in one or more lobes (66% of all presenting cases have single lobe involvement), pleural effusion may be visible in up to 30% of cases. Haemoglobin is often slightly lower than baseline (by a mean drop of 7g/L); leukocytes are often increased.
3.0 Clinical Recommendations for Management of Acute Chest Syndrome or Pneumonia in Sickle Cell Disease

**Inpatient Management**
- Vital sign monitoring
- Refer Vital Sign Monitoring Guidelines

**Administer:**
- Antibiotics as indicated
- Hydration (continuous IV and P0 fluids at maintenance flow rates, increase fluids as needed, if child is dehydrated or inresolvable losses are increased e.g. persistent fever, DO NOT exceed Total Fluid Intake (TFI) of maintenance)
- Analgesics
- Antipyretics
- Bronchodilators (if the child has a history of reactive air disease or wheezing)

**Complete the following if indicated:**
- CBC with reticulocyte counts on admission and then as clinically indicated
- nasopharyngeal swabs (before 1100 H)
- send a swab for mycoplasma PCR if high clinical suspicion for atypical organisms like mycoplasma;
- venous blood gases

**Physiotherapy**
- Refer to Physiotherapy for chest, back, neck or abdomen VDC for mobility and incentive spirometry unless contraindicated
- Refer to Cardiopulmonary Physiotherapy Guidelines for incentive spirometry initiation hours
- Consult may be warranted for patients with history other VDC, and
- Other healthcare professionals should encourage ambulation and activity as per physiotherapists’ recommendation and within child’s tolerance.

**Discuss need for tapping pleural effusion**
- Consult Haematology for worsening hypoxia.

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**ALL children with ACS MUST be admitted to 79CDI under Pediatric Medicine or PICU for critically ill child**
# Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease

## 4.0 References


## 5.0 Related documents

- Fever: Guidelines for Management in Children with Sickle Cell Disease
- Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease
- Fluid and Electrolyte Administration in Children

Attachments:

- [Acute Chest Care Pathway 2021 FINAL.pdf](#)
- [Acute Chest Discharge Criteria Revision 2021.pdf](#)
- [Revision History.docx](#)
- [SC_Clinic Follow Up Revised 2021_FINAL.pdf](#)
- [SCD pain plan_july 2015.pdf](#)