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 sickling)</em></td>
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<tr>
<td>• Pneumococcus</td>
<td>• Hypoventilation secondary to rib/sternal 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>
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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

#### Child with Sickle Cell Disease (SCD) presents to ED with respiratory distress and/or fever (oral or rectal temperature > 38.5°C or axillary temperature > 38°C)

**Complete Initial Assessment and Management:**

1. Refer to ED Fever order set in Epic: complete history, physical exam, and lab investigations.
2. Request chest x-ray if child has a fever, chest pain, tachypnea, or respiratory symptoms.
3. Encourage child to drink orally. If fluid intakes are not tolerated fluid or dehydrated, commence IV 4:1/normal saline at total fluid intake (TFI) of maintenance. See Fluid and Electrolyte Administration in Children.
4. Administer oxygen to maintain O₂ saturation >95%.
5. Start anterolateral (refer to anterolateral for details).
6. Treat with appropriate analgesics and antipyretics (refer to Acute Painful Episodes Vaso-occlusive Crisis and Fever Guidelines for Sickle Cell Disease).
7. Complete rapid flu testing during influenza season to guide use of Tamiflu if clinically indicated.
8. Consult Haematology follow. The fellow must see all seriously ill patients and ensure that all patients are followed by the Sickle Cell team.

**Inpatient Management**

- **Vital signs monitoring:** Refer Vital Signs Monitoring Guidelines
- **Antibiotics:** as indicated
  - Hydroxyzine (continue IV and PO fluids at maintenance flow rates. Increase fluids as needed, if child is dehydrated or insensible losses are increases e.g. persistent fever. DO NOT exceed Total Fluid intake (TFI) of maintenance)
  - Antigens
  - 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
  - Blood culture
  - Nasopharyngeal swab (before 1100 h)
  - Serum Electrolytes (NA, K, glucose, creatinine) should be ordered – refer to Fluid and Electrolyte Guidelines

**History to include:**

- Asthma breathing difficulties
- Fever and infectious symptoms
- Nausea, duration, and severity of pain
- Medications already used
- Family history of sickle cell disease
- Previous episodes of ACS or pneumonias
- Baseline hydration
- Previous episodes of ACS or pneumonias

**Physical exam to include:**

- Vital signs i.e., pulse oximetry and pain score
- Cardiopulmonary
- Hydration status
- Spleen size
- Abdominal distension
- Presence of purpura
- Signs of infection

**Lab investigations to include:**

- CBC, differential and reticulocyte count
- Blood culture
- Continuous O₂ saturation monitoring if Fe or she is in moderate to severe respiratory distress
- Blood type and cross matching (for possible exchange transfusion)
- If respiratory distress
- Blood transfusion laboratory request for cross matching must have SCD diagnosis noted
- Serum Electrolytes (NA, K, glucose, creatinine) should be ordered – refer to Fluid and Electrolyte Guidelines

**ALL children with ACS MUST be admitted to 7ECDE under Pediatric Medicine or PICU for critically ill child

- Inpatient Management
- Vital signs monitoring
- Refer Vital Signs Monitoring Guidelines
- Antibiotics as indicated
  - Hydroxyzine (continue IV and PO fluids at maintenance flow rates. Increase fluids as needed, if child is dehydrated or insensible losses are increases e.g. persistent fever. DO NOT exceed Total Fluid intake (TFI) of maintenance
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**Child discharged home with appropriate follow-up if he/she meets the discharge criteria

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### PRINTABLE VERSION OF CLINICAL PATHWAY

### 4.0 References


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Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease

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