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>
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<tbody>
<tr>
<td><strong>Bacteria</strong></td>
<td>• Pulmonary infarction (in situ sickling)</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><strong>Viruses</strong></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 > 38.5ºC or axillary temperature > 38ºC).

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 not tolerating fluids or dehydrated, commence IV 0.9% normal saline at fluid intake (TFI) of maintenance. See Fluid and Electrolyte Administration in Children.
4. Administer oxygen to maintain O₂ saturation ≥95%.
5. Start antistreptolysin (refer to a hematology for details).
6. Treat with appropriate antibiotics 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 fellow. The fellow must see all seriously ill patients and ensure that all patients are followed by the Sickle Cell team.

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
• Administer
  - Antibiotics as indicated
  - Hydration (continue IV and PO fluids at maintenance fluid rates. Increase fluids as needed, if child is dehydrated or insensible 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
  - Bone marrow if thrombocytopenia
  - Send a swab for mycoplasma PCR (if high clinical suspicion for atypical organisms like mycoplasma; various blood gases

• Physiotherapy:
  - Refer to Physiotherapy for chest, back, neck or abdominal VOC (if mobility and incentive spirometry unless contraindicated)

• Consult may be warranted for patients with limb ischemia or cardiac compromise.

• Discuss need for imaging pleural effusion.
• Consult Haematology for worsening hypoxia.

Physical exam to include:
- Vital signs i.e. pulse oximetry and pain score
- Cardiopulmonary
- Hemoglobin
- Liver/spleen size
- Nuchal rigidity
- Presence of jaundice
- Signs of infection

Lab investigations to include:
- CBC, differential and reticulocyte count
- Blood culture
- Complete O₂ saturation monitoring if fever or child is in moderate to severe respiratory distress
- Blood typing and cross matching (for possible exchange transfusion). If in respiratory distress
- Blood transfusion/institution: request for cross matching must have ACS diagnosis noted
- Serum Electrolytes (NA, K, glucose, creatinine) should be ordered – refer to Fluid and Electrolyte Guidelines

Complete Initial Assessment and Management:

- Serum Electrolytes (NA, K, glucose, creatinine) should be ordered – refer to Fluid and Electrolyte Guidelines

The fellow must see all seriously ill patients and ensure that all patients are followed by the Sickle Cell team.

Child discharged home with appropriate follow-up if he/she meets the discharge criteria.

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