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>
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<tr>
<td>Pneumococcus</td>
<td>Pulmonary infarction (in situ sickling)</td>
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<tr>
<td>Gram-negative bacteria</td>
<td>Hypoventilation secondary to rib/sternal infarction or narcotic administration</td>
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<tr>
<td>Chlamydia pneumoniae</td>
<td>Fat embolism</td>
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<tr>
<td>Mycoplasma pneumoniae</td>
<td>Pulmonary edema secondary to fluid overload</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 HbSß° thalassemia (9.4 events/100 patient-years) or HbSC (5.2 events/100 patient-years); and least often in those with HbSß+ 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.

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

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 not tolerating fluids or dehydrated, commence IV dextrose 5% in 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 antibiotics (refer to antibiotic for details).
7. Complete rapid flu testing during influenza season to guide use of Tamiflu if clinically indicated.
8. Consult may be warranted for patients with limb-threatening pain.
9. Obtain blood cultures:
   - CBC with reticulocyte counts on admission and then as clinically indicated
   - Blood transfusion laboratory request for cross-match
   - Blood type and cross-match for possible exchange transfusion
   - Peripheral blood smear
   - Serum Electrolytes (K, glucose, creatinine) should be ordered – refer to Fluid and Electrolyte Guidelines

Inpatient Management:
- Vital sign monitoring
- Refer Vital Sign Monitoring Guidelines
- Administer:
  - Antibiotics as indicated
  - Hydration: continue IV and PO fluids at maintenance flow rates. Increase fluids as needed. If child is dehydrated or reasonable losses are increases e.g. persistent fever, DO NOT exceed Total Fluid Intake (TFI) of maintenance
  - Analgesics
  - Antipyretics
- Monitor if the child has a history of reactive air disease or wheezing

Lab investigations to include:
- CBC, differential, and reticulocyte count
- Blood culture
- Complete O₂ saturation monitoring if the child is in moderate to severe respiratory distress
- Blood type and cross-match (for possible exchange transfusion), if respiratory distress
- Blood transfusion laboratory request for cross-match

Consult may be warranted for patients with limb-threatening pain.
- Consider exchange transfusion in critically ill child
- Other health care professionals should encourage embolization and activity as per physiotherapists’ recommendation and within child’s tolerance.
- Consult may be warranted for patients with limb-threatening VOD until
- Solar phototherapy

Discuss need for tapping pleural effusion.

Child discharged home with appropriate followup if he/she meets the discharge criteria

SickKids: Guideline for Management of Acute Chest Syndrome (ACS) or Pneumonia

PRINTABLE VERSION OF CLINICAL PATHWAY

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

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References


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:

- SC_clinic follow up.pdf
- discharge criteria_final_apr 1.pdf
- SCD pain plan_july 2015.pdf
- care pathway_final.pdf
- Revision History.docx