Use of this guideline in any setting must be subject to the clinical judgment of those responsible for providing care.

IWK Health

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Acute Chest Syndrome or Pneumonia: Guidelines for Management in Children with Sickle Cell Disease (Adapted from the Hospital for Sick Children)

Version: 1

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.)

Infectious Causes	Non-infectious Causes of ACS
Bacteria • Pneumococcus • Gram-negative bacteria • Chlamydia pneumoniae • Mycoplasma pneumoniae **Viruses** • Respiratory syncytial virus • Para-influenza • Influenza	• Pulmonary infarction (*in situ* sickling) • Hypoventilation secondary to rib/sternal infarction or narcotic administration • Fat embolism • Pulmonary edema secondary to fluid overload

In patients with sickle cell disease, ACS occurs most frequently in patients with hemoglobin genotype SS (12.8 events/100 patient-years); less so in those with HbSß0 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). 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. Hemoglobin 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 > 38.3 ºC or axillary temperature >38ºC)

Complete Initial Assessment and Management:

1. Consult Pediatric Hematology/Oncology and prepare for transfer to IWK.
2. Refer to ED Sickle Cell Fever order set on the APPHON website or at IWK e-access: complete history, physical exam, and lab investigations.
3. Request chest x-ray if child has a fever, chest pain, tachypnea, or respiratory symptoms.
4. Encourage child to drink orally. If not tolerating fluids or dehydrated, commence IV normal saline at total fluid intake (TFI) of 75% of maintenance.
5. Administer oxygen to maintain O2 saturation greater than 95%.
6. Start antimicrobials as per the IWK or APPHON Sickle Cell Fever preprinted orders.
7. Treat with appropriate analgesics and antipyretics (refer to Sickle Cell Pain Admission Orders) and Fever Orders Guidelines for Sickle Cell Disease on the APPHON website or at IWK e-access.
8. If unable to obtain IV access after 2 attempts, consider ultrasound guided IV placement or page anesthesia/anesthesia assistant. Refer to IWK IV policy #1155.
9. Consider simple transfusion in consultation with hematology. Do NOT exceed a post transfusion Hb of 100g/L.
10. Complete rapid flu testing during influenza season to guide use of Tamiflu if clinically indicated.
11. Complete COVID-19 testing to guide antiviral therapy as indicated.

History to include:

- Ascertain breathing difficulties
- Fever and infectious symptoms
- Nature, duration, and severity of pain
- Medications already used
- Associated symptoms
- Previously successful experience with analgesics
- Baseline hemoglobin
- Previous episodes of ACS or pneumonia

Physical exam to include:

- Vital signs including pulse oximetry and pain score
- Cardiopulmonary
- Hydration status
- Spleen size
- Neurologic exam
- Presence of jaundice
- Signs of infection

Monitoring to include:

- Continuous O2 saturation monitoring if he or she is in moderate to severe respiratory distress.

Lab investigations to include:

- CBCD, and reticulocyte count
- Blood culture
- Blood type and screen and cross-matching (for possible exchange transfusion), if in respiratory distress
- Blood transfusion laboratory request for cross-matching must have SCD diagnosis noted.
- Serum Electrolytes (NA, K, glucose, creatinine) should be ordered.

ALL children with ACS **MUST** be admitted to 6-link or ICU for critically ill child.

Inpatient Management

Vital sign Monitoring

- Monitor vital signs every 15 minutes.

Administer:

- Antibiotics as indicated.
- Hydration (continue IV and PO fluids at maintenance flow 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 (Consider consult to acute pain service for hard to manage pain)
- Antipyretics
- Bronchodilators (if the child has a history of reactive air disease or wheezing)

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Complete the following if indicated:

- CBCD with reticulocyte counts on admission and then as clinically indicated.
- nasopharyngeal swab
- send a swab for mycoplasma PCR if high clinical suspicion for atypical organisms like mycoplasma.
- venous blood gases

Physiotherapy:

- Encourage mobility and deep breathing with tolerance.
- Consult physiotherapy for complex mobility needs and/or airway clearance techniques if required.

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

4.0 References

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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