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

Version: 5

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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 haemoglobin 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) (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.

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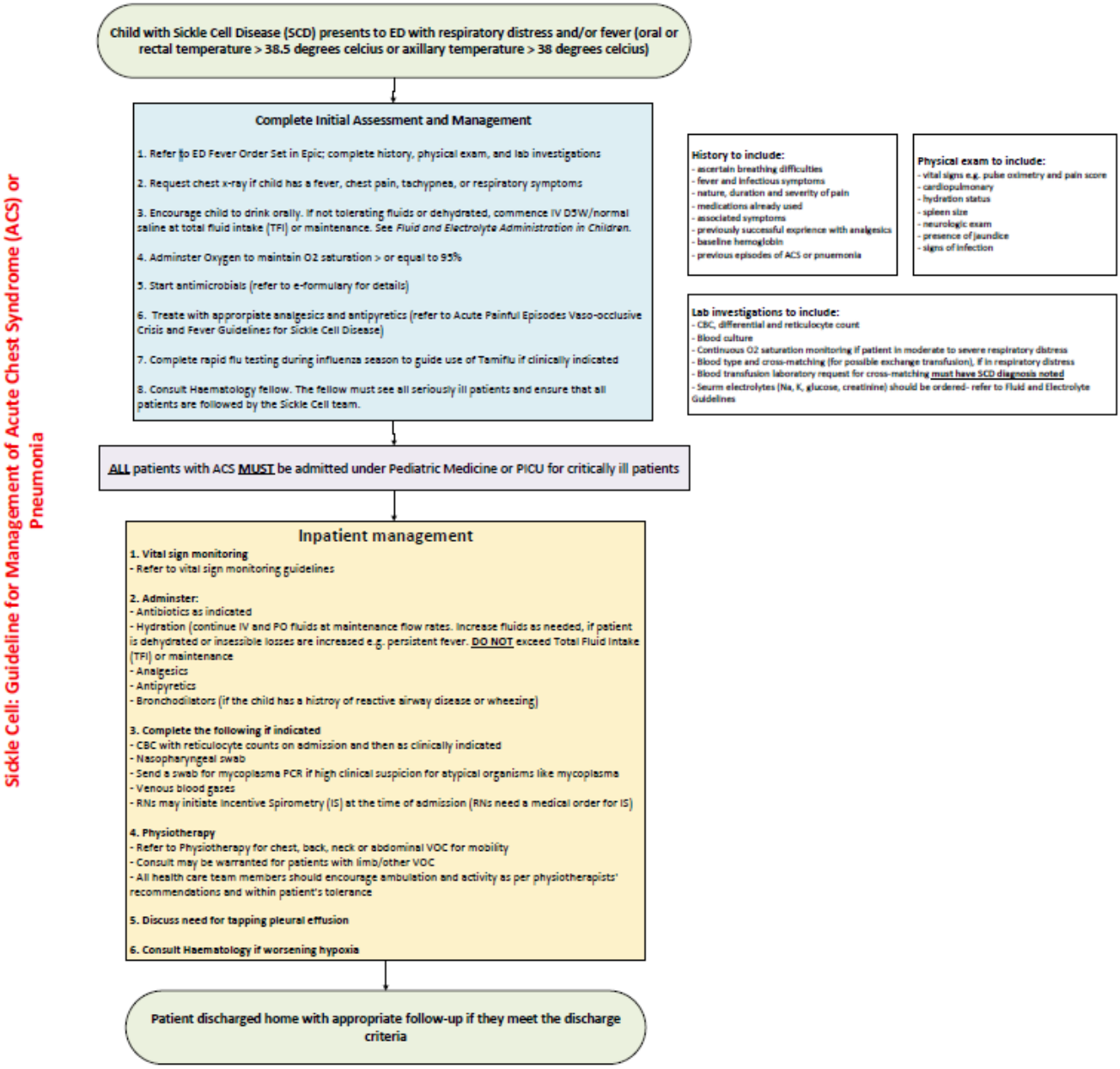
3.0 Clinical Recommendations for Management of Acute Chest Syndrome or Pneumonia in Sickle Cell Disease



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4.0 References

1. Acute complications in children with sickle cell disease: Prevention and management | Canadian Paediatric Society (cps.ca)
2. Castro O, Brambilla DJ, Thorington B, et al. The acute chest syndrome in sickle cell disease: incidence and risk factors. *Blood*. 1994; 84:643–49.
3. Liem RI, O'Gorman MR, Brown DL. Effect of red cell exchange transfusion on plasma levels of inflammatory mediators in sickle cell patients with acute chest syndrome. *Am J Hematol*.2004;76(1):19-25.
4. National Heart, Lung, and Blood Institute. Evidence-based management of sickle cell disease: Expert panel report, 2014.
5. Reid CD, Charache S, Lubin B (eds). *Management and Therapy of Sickle Cell Disease, 3rd edition*. National Institutes of Health Publication No 95-2117, Bethesda, Maryland, 1995.
6. Robieux IC, Kellner JD, Coppes MJ, Shaw D, Brown E, Good C, O'Brodovich H, Manson D, Olivieri NF, Zipursky A, Koren G. Analgesia in children with sickle cell crisis: comparison of intermittent opioids vs. continuous intravenous infusion of morphine and placebo-controlled study of oxygen inhalation. *Pediatr Hematol Oncol*. 1992;9:317–26.
7. Shapiro B. The management of pain in sickle cell disease. *Pediatr Clin North Am*. 1989;36:1029–45.
8. Vichinsky EP, Styles LA, Colangelo LH, Wright EC, Castro O, Nickerson B. Acute chest syndrome in sickle cell disease: clinical presentation and course. *Blood*. 1997;89:1787–92.

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

Acute Chest Discharge Criteria Revision pdf

Revision History.docx

SC_Clinic Follow Up Revised FINAL.pdf

SCD pain plan.pdf

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