Clinical Guideline

Acute Chest Syndrome

Pediatric Emergency, Pediatric Critical Care, & Pediatric Hematology-Oncology

⚠ This guideline serves as a guide and does not replace clinical judgment.

A child with sickle cell disease presenting with:

Respiratory symptoms (cough, chest pain, respiratory distress, dyspnea, or tachypnea) and/or temperature 38.3°C **PLUS** a new pulmonary infiltrate on chest X-ray

Initial Assessment & Evaluation

- Chest X-ray to evaluate for infiltrate
- Physical exam (neuro exam, palpate for spleen)
- Pulse Oximetry
- Blood cultures
- CBC w/diff, Retic count, Type & Screen
- Chemistries including fractioned bilirubin (BMP + liver function panel)
- Respiratory Pathogen panel
- Consult Pediatric Hematology Oncology

Initial Management

Airway

- Maintain oxygen saturations above 92%
- Caution use of corticosteroids due to association of rebound pain and readmission
- Consider inhaled bronchodilators if asthma diagnosis

Pain Control

- Quickly & adequately treat pain to minimize splinting (can worsen ACS)
- See CHoR clinical guidelines or patients individualized pain plan in the problem list in EPIC

Empiric Antibiotics

- Ceftriaxone 50 mg/kg/dose IV/IM q24 hours (max dose 2000mg)
- If RPP positive for *mycoplasma pneumoniae* or *chlamydial pneumoniae*, add azithromycin 10mg/kg (max dose = 500mg) on day 1, then 5mg/kg (max dose = 250mg) q24 hours on days 2-5
- If cephalosporin allergy: levofloxacin
  - Less than 5 years = levofloxacin 10mg/kg q12 hours
  - 5 years of age or older = levofloxacin 10mg/kg q24 hours (max dose = 750mg)
- If acutely ill with large/progressive pulmonary infiltrates and/or is a known MRSA carrier consider empiric vancomycin therapy

Fluid Status

- Maintain euvolemia (do not hyperhydrate)
- If continuous IV fluids indicated, max oral intake + IV fluids should be equal to 1x maintenance needs
- Consider IV furosemide (0.5-1mg/kg) if signs of fluids overload present

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Blood Products

- Consider simple vs. exchange transfusion for severe or worsening symptoms – discuss with pediatric hematology oncology fellow or attending
- All blood products should be sickle-cell negative, leukoreduced, fully cross-matched if possible
- Pre-medicate only if history of transfusion reaction (e.g. acetaminophen + diphenhydramine)
- Note: delayed hemolytic transfusion reaction may present 1 week later

Simple Transfusion

- Consider in patients:
  - Hb ≥2g/dL below baseline
  - Hypoxia
- If on hydroxyurea, obtain %HbS ("Hemoglobin Fract/Quant") pre-transfusion if not obtained in the previous 3 months
- Amount: Refer to Transfusion Guidelines in Resident Manual
- Do not exceed hemoglobin >10g/dL or hematocrit >30%

Exchange Transfusion

- Consider in patients:
  - Patient not sufficiently anemic for simple transfusion
  - Persistent hypoxia
  - Requires mechanical ventilation
  - Unresponsive to simple transfusion
- Obtain %HbS ("Hemoglobin Fract/Quant") pre- and post-exchange (call Path resident to request STAT results)
- If desire to exchange: notify pediatric hematology oncology team, blood bank, and PICU
- Goal: HbS <30% while not exceeding Hb 10g/dL
- Line placement in PICU (stiff-walled dialysis catheter)

Inpatient Management

- Daily CBC with diff and reticulocyte count
- Vitals q2-4 hours (including SpO2 and Pain Scale)
- Strict input and output
- Daily weights
- Continuous pulse oximetry with goal to maintain >95% oxygen saturation
- Incentive spirometry (monitor compliance)
- Consult peds pulmonology
- Encourage ambulation at least twice daily when clinically appropriate and consult physical therapy (if specific need)
- Consider vigorous chest physiotherapy
- Monitor for signs of neurologic complications
- Repeat chest x-ray in the setting of respiratory or clinical changes
- Continue ceftriaxone (and hold home penicillin prophylaxis when receiving broad-spectrum antibiotics) and continue azithromycin if RPP positive for *Mycoplasma pneumoniae* or *Chlamydia pneumoniae*

Discharge Criteria & Follow-Up

- Hemoglobin stabilized
- Oxygenation stable on room air
- Afebrile for greater than 24 hours
- Tolerating oral medications, adequate oral fluid intake, and pain control
- Antibiotics to complete 7-day course
- For severe cases of acute chest syndrome: Follow up in 1-2 weeks in pediatric hematology oncology clinic
- For mild to moderate cases: Follow up at regular scheduled visit
- Referral or follow up with pediatric pulmonology per consult recommendations

Executive Summary

Acute Chest Syndrome

Pediatric Emergency, Pediatric Critical Care, & Pediatric Hematology-Oncology

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References

Howard J, Hart N, Roberts-Harewood M, et al. Guideline on the management of acute chest syndrome in sickle cell disease. *Br J Haematol*. 2015;169(4):492-505. doi:10.1111/bjh.13348

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Retrieval website: **https://www.chrichmond.org/health-care-professionals/chor-clinical-guidelines**

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