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Pediatric Suspected Acute Chest Syndrome Pathway

Arkansas Children's
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Purpose

To standardize the care of sickle cell disease patients who present to Arkansas Children's with acute chest syndrome

Background

Acute chest syndrome is defined as an acute illness with fever and/or respiratory symptoms in the setting of sickle cell disease (SCD), accompanied by a new lung infiltrate on chest x-ray that is consistent with alveolar consolidation, but not atelectasis, involving at least one entire lung segment [1,2].

Acute chest syndrome (ACS) is the second most common cause of hospitalization and is the most common cause of admission to intensive care units and death in patients with SCD [1]. It is also a frequent complication in patients admitted with sickle cell vaso-occlusive crisis [3]. ACS is a form of acute lung injury in SCD. The three major causes of ACS that have been hypothesized include: pulmonary infections, embolization of bone marrow fat, and pulmonary infarction. A vicious cycle arises where lung injury results in ventilation-perfusion mismatch and hypoxemia, which then leads to increased deoxygenation of hemoglobin S, with resulting hemoglobin polymerization with erythrocyte sickling, followed by bone marrow infarction and pulmonary vaso-occlusion [1,3].

Patients with SCD can present with ACS, or ACS may develop at some point after the onset of a pain crisis. Therefore, vigilance should be maintained whenever a patient has a sickle cell pain crisis [2]. Patients with ACS should also be monitored closely for risk factors for a more severe clinical course, which include worsening hypoxemia, increasing respiratory rate, increasing work of breathing, decreasing platelet count, decreasing hemoglobin concentration, or the involvement of multiple lobes of the lung on chest x-ray [2].

Clinical management focuses on supportive care, close monitoring for clinical deterioration, optimal fluid management, frequent respiratory therapies, appropriate antibiotics, adequate analgesia, and blood transfusions as indicated.

Pediatric Suspected Acute Chest Syndrome - ED Pathway

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Hold PCN prophylaxis while on broad-spectrum antibiotics

Inclusion Criteria:

New lung infiltrate in patient with sickle cell disease (NOT atelectasis)
AND
At least one of the following:

- Temp ≥ 38.3 °C
- Chest pain
- Cough
- Wheeze
- Tachypnea
- Increased work of breathing
- Hypoxemia
- Crackles

Perform physical exam and complete the following:

Labs to obtain:

- CBC with differential
- Retic
- Type and cross
- Complete metabolic panel (CMP)
- Blood culture if febrile (Temp ≥ 38.3°C) / urine culture if symptomatic
- Consider respiratory viral panel
- Consider venous blood gas

Medications to administer:

- Initiate pain medications per medication dosing table (pg. 2) if needed
- Initiate antibiotics – Ceftriaxone/Azithromycin (if cephalosporin allergy consider vancomycin and/or levofloxacin)

Hem/Onc Service Admit

- Hem/Onc consult for ACNW admissions
- IV fluids to maintain euvolemia – ¾ to 1x maintenance or less if good po intake
- Oxygen saturation goal > 92%
- Assess if PRBC transfusion* needed

ICU Admit Criteria:

- Need for oxygen supplementation >15L via HFNC
- Need for ventilation based on signs of respiratory failure or an abnormal blood gas
- Need for exchange transfusion/apheresis
- Change in neurological status
- Clinical signs of sepsis

Does patient require exchange transfusion?

YES

Exchange Transfusion Criteria:

- Rapid worsening in respiratory status
- Need for oxygen supplementation >15L via HFNC
- Impending/existing respiratory failure
- Neurologic/mental status change
- Avoid hyper-viscosity associated with a simple transfusion
  - Patients with HbSC and HbSß + thalassemia who have baseline HgB levels of ≥ 9
- **Post-exchange goal: HgbS < 30%**

NO

PRBC Transfusion Criteria:

- Decrease in hemoglobin 1-2 g/dL from baseline
- Hypoxemia
- High cardiac output (persistent tachycardia)
- Symptoms (lightheadedness, easy fatigability)

Recommendations (based on current hemoglobin):

- 10-15 ml/kg packed RBCs for infants and children
- 1-2 units packed RBCs for adults

All PRBCs must be HbS negative, leukoreduced, antigen matched
DO NOT transfuse to over Hgb 10 g/dL

Pediatric Suspected Acute Chest Syndrome - Inpatient Pathway

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Hold PCN prophylaxis while on broad-spectrum antibiotics

Inclusion Criteria:

New lung infiltrate in patient with sickle cell disease (NOT atelectasis)
AND
At least one of the following:

- Temp ≥ 38.3 °C
- Chest pain
- Cough
- Wheeze
- Tachypnea
- Increased work of breathing
- Hypoxemia
- Crackles

Perform physical exam and complete the following:

Daily labs:

- CBC with differential
- Retic

Medications to administer:

- Pain medications per medication dosing table (pg. 2)
- Continue antibiotics – Ceftriaxone/Azithromycin (if cephalosporin allergy consider vancomycin and/or levofloxacin)

Upon admission:

- Hem/Onc consult for ACNW admissions
- Vital signs Q4 hours minimum with continuous pulse oximetry
- Strict I/O
- Incentive spirometry Q2 hours while awake (with vitals 2200-0800)
- Titrate respiratory support (low threshold for HFNC, CPAP, NIPPV)
- Oxygen saturation goal >92%
- Assess if PRBC transfusion* needed
- Chest physiotherapy Q4 hours

ICU/transfer to higher level of care criteria:

- Need for oxygen supplementation >15L via HFNC
- Need for ventilation based on signs of respiratory failure or an abnormal blood gas
- Need for exchange transfusion/apheresis
- Change in neurological status
- Clinical signs of sepsis

Exchange Transfusion Criteria:

- Rapid worsening in respiratory status
- Need for oxygen supplementation >15L via HFNC
- Impending/existing respiratory failure
- Neurologic/mental status change
- Clinical concerns for sepsis
- Avoid hyperviscosity associated with a simple transfusion
  - Patients with HbSC and HbSß + thalassemia who have baseline HgB levels of ≥ 9

Post-exchange goal: HgbS < 30%

YES ← Does patient require exchange transfusion? → **NO**

PRBC Transfusion Criteria:

- Decrease in hemoglobin 1-2 g/dL from baseline
- Hypoxemia
- High cardiac output (persistent tachycardia)
- Symptoms (lightheadedness, easy fatigability)

Recommendations (based on current hemoglobin):

- 10-15 ml/kg packed RBCs for infants and children
- 1-2 units packed RBCs for adults

All PRBCs must be HbS negative, leukoreduced, antigen matched
DO NOT transfuse to over Hgb 10 g/dL

Discharge Criteria:

- Back to respiratory baseline
- Afebrile for at least 24 hours
- Negative blood culture ≥ 48 hours
- Good PO/enteral intake
- Pain controlled with PO pain medications only
- Outpatient incentive spirometry plan in place
- Hgb and retic stable
- Follow-up with Hem/Onc Sickle Cell/Sickle Cell Pulmonary clinic within 4 weeks
- Additional PO antibiotics as needed to complete 7 day course (**click here**)

Inpatient Medication Dosing

HOLD PCN prophylaxis while on broad-spectrum antibiotics

Medication	Route	Dose
Ceftriaxone (pneumococcal coverage)	IV	75 mg/kg/day every 24 hours (max 2 grams/day)
Levofloxacin (if cephalosporin allergy)	PO/IV	<5 years - 10 mg/kg/dose BID
		≥5 years – 10 mg/kg/dose once daily (max 750 mg/day)
Azithromycin (atypical coverage)	PO/IV	10 mg/kg on day 1 (max 500 mg/day), followed by 5 mg/kg/day once daily on days 2-5 (max 250 mg/day)
Vancomycin (therapeutic drug monitoring required)	IV	60 mg/kg/day divided every 6-8 hours (consider using previous dosing if appropriate)
Acetaminophen	PO/IV	15 mg/kg PO Q6 hours PRN fever
Albuterol	Inhalation	Per beta care pathway
Toradol (only for ≥ 2 years of age)	IV	0.5 mg/kg IV Q6 hours scheduled for 72 hours
Oxycodone	PO	0.1 mg/kg Q4 hours PRN moderate pain (max 5 mg/dose, consider up to 10mg for opioid tolerance)
Morphine	IV	0.1 mg/kg Q2-4 hours PRN severe pain (max 5 mg/dose, consider up to 10mg for opioid tolerance)

Discharge Medication Dosing

Resume PCN prophylaxis at discharge

Medication	Route	Dose
Amoxicillin Clavulanate	PO	Amoxicillin component-90 mg/kg/day in 2 divided doses (max 4 grams/day)
Azithromycin (if not completed inpatient)	PO	10 mg/kg on day 1 (max 500 mg/day), followed by 5 mg/kg/day once daily on days 2-5 (max 250 mg/day)
Cefpodoxime infants >3 months to children <12 years	PO	10 mg/kg/day divided every 12 hours (max 400 mg/day)
Cefpodoxime children ≥12 years	PO	200 mg every 12 hours (max 400 mg/day)
Cefuroxime	PO (tablet formulation **ONLY**)	<30 kg 250 mg BID (max 500 mg/day)
		≥30 kg 500 mg BID (max 1 gram/day)
Levofloxacin	PO	<5 years - 10 mg/kg/dose BID
		≥5 years – 10 mg/kg/dose once daily (max 750 mg/day)

Metrics

1. Time to first antibiotic (goal < 60 minutes)
2. Order set utilization
3. Length of stay
4. Readmission rate
5. Rate of emergent escalations

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