Sickle Cell Disease: Acute Evaluation and Management - Pediatric - Emergency Department Clinical Practice Guideline

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Executive Summary

Guideline Overview

Implementation of clinical algorithms and guidelines for pediatric Sickle Cell Disease have been shown to promote more rapid stabilization and reduce the use of inappropriate therapies.[1-3] The following guideline outlines care which should be provided in the emergency department.

Key Practice Recommendations

Vaso-occlusive Crisis (VOC)

1. Perform a pain assessment (e.g., determine severity, location, characteristics, associated symptoms) based on patient self-report and observation.[4] *(HHS Consensus- Adapted)*
2. Rapidly assess recent analgesic use (opioid or non-opioid) *(HHS Consensus- Adapted)* and initiate analgesic therapy.[4,5] Use of intranasal fentanyl (1.5 mcg/kg; max dose 100 mcg/dose) is recommended until IV access is established.[6] *(UW Health High quality evidence, strong recommendation)*
3. Base analgesic selection on pain assessment, associated symptoms, outpatient analgesic use, patient knowledge of effective agents and doses, and past experiences with side effects.[4] *(HHS Consensus-Adapted)* Providers should reference record of historical therapy that the patient has received and/or the individualized pain plan documented in the medical record.[5]
4. Perform regular assessment for side effects and/or excessive sedation.[4] *(UW Health Moderate quality evidence, strong recommendation)*
5. All patients who do not achieve adequate pain relief should be admitted to the hospital for additional therapy.[7]

Fever

1. Immediately evaluate the patient with a history and physical, complete blood count with differential, reticulocyte count, and blood culture.[4,8] A urine culture may be obtained if a urinary tract infection is suspected.[4] *(HHS Consensus- Panel Expertise)*
2. Promptly administer ongoing empiric parenteral antibiotics that provider coverage against *Streptococcus pneumoniae* and gram-negative enteric organisms.[4] *(HHS Consensus- Panel Expertise)*
3. Treat patients with SCD who have ACS with an intravenous cephalosporin, an oral macrolide antibiotic, supplemental oxygen (to maintain oxygen saturation > 95%) and close monitoring for bronchospasm, acute anemia, and hypoxemia.[4,8] *(HHS Low quality evidence, strong recommendation)*
4. Patients who appear ill should be hospitalized for close observation and intravenous antibiotic therapy.[4] *(HHS Consensus- Panel Expertise)* However, patients who lack certain high risk factors (i.e., white blood cell count < 5,000/mm^3 or > 30,000/mm^3, fever > 40°C, or "ill-appearing") may be managed as an outpatient following administration of intravenous empiric, antipneumococcal antibiotic (e.g., ceftriaxone).[8] *(UW Health Very low quality evidence, weak/conditional recommendation)*

Companion Documents

1. Evaluation and Initial Management of Children with Sickle Cell Disease and Pain Suspected to be a Vaso-Occlusive Event Algorithm
2. Evaluation and Initial Management of Children with Sickle Cell Disease and Fever Algorithm

Scope

Disease/Condition(s): Sickle Cell Disease (SCD)

Clinical Specialty: Pediatric Emergency Medicine, Emergency Medicine, Pediatric Hematology/Oncology, Pediatric Intensive Care

Intended Users: Physicians, Advanced Practice Providers, Registered Nurses, Pharmacists

Objective(s): To outline evidence-based recommendations for the evaluation and management of patients presenting to the emergency department with an acute Sickle Cell Disease complication.

Target Population: Pediatric patients with Sickle Cell Disease.

Methodology

Methods Used to Collect/Select the Evidence:
Electronic database searches (e.g., PUBMED) were conducted by the guideline author(s) and workgroup members to collect evidence for review. Expert opinion and clinical experience were also considered during discussions of the evidence.

Methods Used to Formulate the Recommendations:
The workgroup members agreed to adopt recommendations developed by external organizations and/or arrived at a consensus through discussion of the literature and expert experience. All recommendations endorsed or developed by the guideline workgroup were reviewed and approved by other stakeholders or committees (as appropriate).

Methods Used to Assess the Quality of the Evidence/Strength of the Recommendations:
Recommendations developed by external organizations maintained the evidence grade assigned within the original source document and were adopted for use at UW Health.

Internally developed recommendations, or those adopted from external sources without an assigned evidence grade, were evaluated by the guideline workgroup using an algorithm adapted from the Grading of Recommendations Assessment, Development and Evaluation (GRADE) methodology (see **Figure 1** in Appendix A).

Rating Scheme for the Strength of the Evidence/Recommendations:
See Appendix A for the rating scheme(s) used within this document.

Recognition of Potential Health Care Disparities: Patients presenting to the emergency department with Sickle Cell Disease experienced 25-50% longer wait times to see a physician after arrival than other patients with long bone fracture or other conditions.[9] Providers should triage and treat this patient population quickly, especially as many often present with higher level of pain than the general population.

Introduction

Vaso-occlusive crisis (VOC) and infection are the two most common reasons for hospitalization in patients with Sickle Cell Disease. Implementation of clinical algorithms and guidelines for pediatric Sickle Cell Disease have been shown to promote more rapid stabilization and reduce

the use of inappropriate therapies.[1-3] The following guideline outlines the care which should be provided in the emergency department for pediatric patients with Sickle Cell Disease.

Recommendations

Management of a Vaso-Occlusive Crisis (VOC)

1. The initial medical assessment should focus on detection of the following medical complications requiring specific therapy: infection, dehydration, acute chest syndrome (fever, tachypnea, chest pain, hypoxia, and/or rales), or severe anemia.[5] In patients with an oxygen saturation < 95% on room air, administer oxygen.[4] *(HHS Consensus- Panel Expertise)*
2. Perform a pain screen and assessment (e.g., determine severity, location, quality, and frequency) based on patient self-report and observation.[4] *(HHS Consensus- Adapted)*
    a. If a pain rating scale is used, it should be chosen based upon patient age and cognitive ability. *(UW Health Moderate quality evidence, strong recommendation)* The Numeric Pain Scale[10] is recommended in patients age 8 years or older, the Pain Face Scale-Revised (FPS-R)[11] in patients age 4-8 years, and the Neonatal/Infant Pain Scale (NIPS)[12] should be used in patients age less than 1 year. The Face Legs Activity Cry and Consolability Scale- Revised (FLACC-R)[13] may be used in patients ages 1 year and above who are unable to verbally report pain due to cognitive or developmental delays.
    b. Patients are often able to recognize whether the pain is typical of their Sickle Cell Disease.[5] If the VOC pain is atypical per patient report, providers should investigate other possible etiologies of pain.[4,5] *(HHS Consensus- Adapted)*
3. Rapidly assess recent analgesic use (opioid or non-opioid) *(HHS Consensus- Adapted)* and initiate analgesic therapy.[4,5] Use of intranasal fentanyl (1.5 mcg/kg; max dose 100 mcg/dose) is recommended until IV access is established.[6] *(UW Health High quality evidence, strong recommendation)*
4. Once IV access is established, it is important to obtain a complete blood count with differential, reticulocyte count and Type and Screen. *(UW Health Very low quality evidence, strong recommendation)*. A blood culture is suggested if a fever is present. *(UW Health Very low quality evidence, weak/conditional recommendation)*
5. In euvolemic patients who are unable to drink fluids, provide intravenous hydration at no more than the maintenance rate to avoid over-hydration.[4,7] *(HHS Consensus- Adapted)*
6. There are no large controlled studies of analgesic regimens in Sickle Cell Disease and a number of smaller studies have failed to define any optimal regimens.[4,5] Base analgesic selection on pain assessment, associated symptoms, outpatient analgesic use, patient knowledge of effective agents and doses, and past experiences with side effects.[4] *(HHS Consensus-Adapted)* Providers should reference record of historical therapy that the patient has received and/or the individualized pain plan documented in the medical record.[5]
7. In patients with mild to moderate pain who report relief with NSAIDs in the absence of contraindications, continue treatment with NSAIDS.[4] *(HHS Low quality evidence, moderate recommendation)* A low dose opioid should also be administered in conjunction with the NSAID.[5,8] *(UW Health Low quality evidence, weak/conditional recommendation)*

8. In patients with severe pain, rapidly initiate treatment with parenteral opioids.[4] *(HHS High quality evidence, strong recommendation)* NSAIDs should also be administered, if not taken in the last 6 hours.[5,8] *(UW Health Low quality evidence, weak/conditional recommendation)*
    a. Reassess pain and re-administer opioids if necessary for continued severe pain every 15-30 minutes until pain is under control.[4] *(HHS Consensus-Adapted)*
    b. Consider maintenance or dose escalation by 50% based upon response to analgesic therapy until pain is controlled.[4] *(UW Health Very low quality evidence, weak/conditional recommendation)*
9. Perform regular assessment for side effects and/or excessive sedation.[4] *(UW Health Moderate quality evidence, strong recommendation)*
10. Use of non-pharmacotherapeutic interventions (e.g., local heat application, distraction) is recommended as adjunctive therapy.[4] *(HHS Consensus- Adapted)*
11. All patients who do not achieve adequate pain relief should be admitted to the hospital for additional therapy.[7] Opioid administration by patient-controlled analgesia (PCA) is preferred over as requested (PRN) administration.[4] *(HHS Low quality evidence, moderate recommendation)* Providers may consider initiating a PCA in the emergency department while awaiting inpatient admission. For dosing recommendations and consideration of side effects, refer to the following resources:
    a. Pain Care Fast Fact: IV Patient Controlled Analgesia in the Pediatric Patient
    b. UW Health Continuous Opioid Therapy – Adult/Pediatric – Inpatient Clinical Practice Guideline.
12. Prior to discharge from the emergency department, patients may be educated on techniques for VOC prevention including avoiding triggers (e.g., infection, emotional stress, exposure to cold, wind, and high altitude), use of hydroxyurea, and maintenance of intravascular volume via oral fluid intake.[8] *(UW Health Very low quality evidence, weak/conditional recommendation)*
13. Red blood cell transfusion is not recommended to resolve VOC.[4,8] *(UW Health Low quality evidence, strong recommendation)*

Management of SCD Patients with a Fever (≥ 101.3°F or ≥ 38.5°C)

1. Febrile illnesses in patients with Sickle Cell Disease are considered an emergency due to the possibility of penicillin-resistant organisms and incomplete vaccination status, despite the declining incidence of invasive pneumococcal infection as a result of prophylactic penicillin and pneumococcal vaccination.[4] Immediately evaluate the patient with a history and physical, complete blood count with differential, reticulocyte count, and blood culture.[4,8] A urine culture and urinalysis may be obtained if a urinary tract infection is suspected.[4,7] *(HHS Consensus- Panel Expertise)*

    a. Providers may confirm the patient's immunization status, as this information can aid in the development of differential diagnoses.[7] *(UW Health Low quality evidence, weak/conditional recommendation)*

    b. Additional laboratory tests to consider for patients in pain with a fever include a Type and screen, potassium, sodium, bicarbonate, and chloride. *(UW Health Low quality evidence, weak/conditional recommendation)*

2. Patients who appear ill should be hospitalized for close observation and intravenous antibiotic therapy.[4] *(HHS Consensus- Panel Expertise)*

3. Promptly administer ongoing empiric parenteral antibiotics that provider coverage against *Streptococcus pneumoniae* and gram-negative enteric organisms.[4] *(HHS Consensus- Panel Expertise)*

4. Acute chest syndrome (ACS) is a common and potentially lethal complication of Sickle Cell Disease.[8] The incidence is highest (25 episodes/100 patient years) in children between the ages of 2 and 5 years.[8] In patients whose febrile illness is accompanied by shortness of breath, tachypnea, cough, and/or rales, it is recommend to obtain an immediate chest x-ray to investigate for acute chest syndrome (ACS).[4] *(HHS Consensus- Panel Expertise)* ACS is defined by a new pulmonary infiltrate on chest x-ray in addition to one or more of the following: fever, tachypnea, dyspnea, hypoxia, and chest pain.[7,8]

    a. Treat patients with SCD who have ACS with an intravenous cephalosporin, an oral macrolide antibiotic, supplemental oxygen (to maintain oxygen saturation > 95%) and close monitoring for bronchospasm, acute anemia, and hypoxemia.[4,8] *(HHS Low quality evidence, strong recommendation)*

    b. Hospitalize patients with ACS.[4] *(HHS Consensus- Panel Expertise)* Patients may be admitted to the PICU or general care floor under the Pediatric Hematology/Oncology service based upon their supplemental oxygen needs.

5. Patients who lack certain high risk factors (i.e., white blood cell count < 5,000/mm^3 or > 30,000/mm^3, fever > 40°C, or "ill-appearing") may be managed as an outpatient following administration of intravenous empiric, antipneumococcal antibiotic (e.g., ceftriaxone).[8] *(UW Health Very low quality evidence, weak/conditional recommendation)* When deciding whether to discharge a patient from the emergency department, it is also important to consider the patient's age, ability of the family to return promptly for recurrent fever or clinical deterioration, and the availability of close follow-up.[8]

6. In febrile patients who have localized or multifocal bone tenderness, especially when accompanied by erythema and swelling, include bacterial osteomyelitis in the differential diagnosis and manage accordingly.[4] *(HHS Consensus- Panel Expertise)*

UW Health Implementation

Potential Benefits:

- Pain relief
- Treatment of infection

Potential Harms:

- Oversedation

UW Health Policies & Procedures

1. UW Health Clinical Policy 3.5.5- Pain Management
2. UW Health Clinical Policy 6.1.2- Intravenous Patient Controlled Analgesia (PCA) for Adult and Pediatric Patients

Patient Resources

1. HFFY #7267- A Guide for Families of Children with Sickle Cell Anemia at AFCH
2. HFFY #4273- Intravenous Patient Controlled Analgesia (IV PCA)
3. HFFY #7233- Intranasal Fentanyl Frequently Asked Questions
4. HFFY #6835- Non-Drug Pain Control for Kids
5. HFFY #7711- Using Pediatric Pain Scales: Neonatal Infant Pain Scale (NIPS)
6. HFFY #7712- Using Pediatric Pain Scales: Faces Legs Activity Cry Consolability Revised Scale (FLACC-R)
7. HFFY #7713- Using Pediatric Pain Scales: Faces Pain Scale
8. HFFY #7714- Using Pediatric Pain Scales: Numeric Rating Scale
9. Kids Health- Sickle Cell Disease
10. Kids Health- Sickle Cell Crisis (Pain Crisis)

Guideline Metrics

1. Time to IN fentanyl administration in VOC patients
2. Time to administration of antibiotics in patients presenting with a fever
3. Adherence to recommended antibiotic regimens

Implementation Plan/Clinical Tools

1. Guideline will be posted on uConnect in a dedicated location for Clinical Practice Guidelines.
2. Release of the guideline will be advertised in the Physician/APP Briefing newsletter.
3. Content and hyperlinks within clinical tools, documents, or Health Link related to the guideline recommendations (such as the following) will be reviewed for consistency and modified as appropriate.

UW Health Clinical Practice Guidelines
Influenza and Pneumococcal Vaccination – Adult/Pediatric – Inpatient/Ambulatory
Continuous Opioid Therapy – Adult/Pediatric – Inpatient

Delegation Protocols
Immunization – Adult/Pediatric – Inpatient [60]
Emergency Department Immediate Orders – Adult/Pediatric [61]

Order Sets & Smart Sets
ED – Sickle Cell Disease with Pain/Fever – Pediatric [5752]
ED – Immediate Orders Delegation Protocol – RN/ED Tech – Pediatric [4274]

Disclaimer

Clinical practice guidelines and clinical reference tools assist clinicians by providing a framework for the evaluation and treatment of patients. This tool outlines the preferred approach for most patients. It is not intended to replace a clinician's judgment or to establish a protocol for all patients. It is understood that some patients will not fit the clinical condition contemplated and that a clinical reference tool or clinical practice guideline will rarely establish the only appropriate approach to a problem.

References

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2. Kavanagh PL, Sprinz PG, Wolfgang TL, et al. Improving the Management of Vaso-Occlusive Episodes in the Pediatric Emergency Department. *Pediatrics.* 2015;136(4):e1016-1025.
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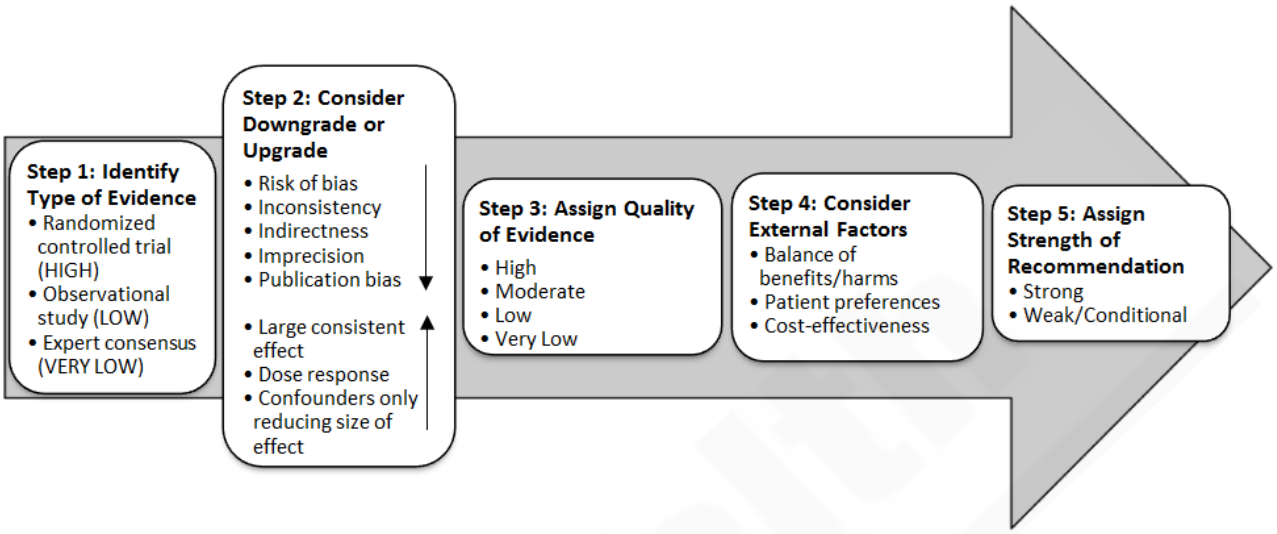
13. Voepel-Lewis T, Zanotti J, Dammeyer JA, Merkel S. Reliability and validity of the face, legs, activity, cry, consolability behavioral tool in assessing acute pain in critically ill patients. *Am J Crit Care.* 2010;19(1):55-61; quiz 62.

Appendix A. Evidence Grading Scheme(s)

Figure 1. GRADE Methodology adapted by UW Health



GRADE Ranking of Evidence

High	We are confident that the effect in the study reflects the actual effect.
Moderate	We are quite confident that the effect in the study is close to the true effect, but it is also possible it is substantially different.
Low	The true effect may differ significantly from the estimate.
Very Low	The true effect is likely to be substantially different from the estimated effect.

GRADE Ratings for Recommendations For or Against Practice

Strong	The net benefit of the treatment is clear, patient values and circumstances are unlikely to affect the decision.
Weak/conditional	Recommendation may be conditional upon patient values and preferences, the resources available, or the setting in which the intervention will be implemented.

Evaluation and Initial Management of Children with Sickle Cell Disease and Pain Suspected to be a Vaso-Occlusive Event – Pediatric – Emergency Dept. Algorithm

UW Health

Patient Population:
HbSS, SC, $S\beta^0$-thalassemia

Patient Presentation

- Perform cardiorespiratory monitoring, pulse oximetry, and pain screen/assessment
- Administer O_2 if O_2 saturation < 95%
- Administer IN fentanyl 1.5 mcg/kg (max 100 mcg/dose) ASAP (until IV access established)
- Obtain initial labs: CBC with differential, reticulocyte count, Type and Screen

- Give normal saline bolus 20 mL/kg (max 1 L) as indicated
- Consider 2 view chest x-ray if tachypneic, chest pain, shortness of breath and/or rales
- If fever, obtain blood culture and refer to Evaluation and Initial Management of Children with Sickle Cell Disease and Fever – Pediatric – Emergency Dept. Algorithm

Reference historical therapy received and/or pain plan documented in the patient's medical record (Problem List)

Is pain well controlled?

Yes → Administer PO oxycodone + PO ibuprofen (if no NSAID in last 6 hours) ***REASSESS IN 60 MINUTES***

Is pain well controlled?

No → Administer IV opioid + IV ketorolac (if no NSAID in the last 6 hours) ***REASSESS IN 20 MINUTES***

Is pain well controlled?

No → IV opioid: 50% loading dose ***REASSESS IN 20 MINUTES***

Is pain well controlled?

Yes → Contact Peds Hem/Onc to discuss possible discharge and follow-up

Meet discharge criteria?

No → ADMIT TO PEDS HEM/ONC

Yes → DISCHARGE
Provide prescription for 3 days of scheduled oxycodone, ibuprofen, and miralax.

Is pain well controlled?

Yes → Contact Peds Hem/Onc to discuss possible discharge and follow-up

No → IV opioid: 50% loading dose

ADMIT TO PEDS/HEM ONC
- Contact Peds Hem/Onc with lab results and patient status
- Consider initiation of PCA or scheduled long-acting oral opioid.

Discharge Considerations
- Pain well-controlled
- Tolerating PO liquids
- Absence of other acute SCD complications

It is preferred to base dosing off of successful historical therapy. If prior history unavailable, consider standard dosing below.

NSAID	Loading Dose
Ibuprofen PO	10 mg/kg/dose (max 600 mg/dose)
Ketorolac IV	0.5 mg/kg/dose (max 30 mg/dose)
Opioid	**Loading Dose**
Fentanyl IV	1 mcg/kg (max 100 mcg/dose)
Morphine IV	0.1-0.15 mg/kg/dose (max 10 mg/dose)
Hydromorphone IV	0.02-0.03 mg/kg/dose (max 2 mg/dose)
Oxycodone PO	0.1-0.2 mg/kg (max 10 mg for initial dose)

References:
1. National Heart L, and Blood Institute. Evidence-based Management of Sickle Cell Disease. Expert Panel Report: U.S. Department of Health and Human Services National Institutes of Health; 2014:32-38.
2. Rees DC, Olujohungbe AD, Parker NE, et al. Guidelines for the management of the acute painful crisis in sickle cell disease. Br J Haematol. 2003;120(5):744-752.
3. Fein DM, Avner JR, Scharbach K, Manwani D, Khine H. Intranasal fentanyl for initial treatment of vaso-occlusive crisis in sickle cell

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Evaluation and Initial Management of Children with Sickle Cell Disease and Fever – Pediatric – Emergency Dept. Algorithm

INTERVENE IMMEDIATELY- PATIENT MAY BE CRITICALLY ILL

Patient Population:
Age > 2 months and ≤ 18 years
HbSS, SC, $S\beta^0$-thalassemia
Fever ≥ 38.5°C in the past 48 hours

Patient Presentation

- Perform cardiorespiratory monitoring, pulse oximetry, and pain screen/assessment
- Administer oxygen if O_2 saturation < 95%
- Initial labs: CBC with differential, reticulocyte count, blood culture
- Consider immediate chest x-ray if tachypneic, chest pain, shortness of breath and/or rales
- Consider additional labs such as: total bilirubin, creatinine, urinalysis and urine culture. If in pain, consider Type and Screen, potassium, sodium, bicarbonate, and chloride and refer to Evaluation and Initial Management of Children with Sickle Cell Disease and Pain Suspected to be a Vaso-Occlusive Event – Pediatric – Emergency Dept. Algorithm
- Check immunization status
- Start maintenance fluids (bolus if clinically dehydrated)

***Historical Red Flags**

- History of Acute Chest Syndrome
- History of stroke
- History of splenic sequestration
- History of sepsis or bacteremia
- Reactive airway disease
- Requires chronic transfusions
- History of PICU admission for SCD related complications

Is the pt ill appearing or any historical red flags*?

- Yes → Acute chest syndrome?**
  - No → Administer IV ceftriaxone → Consult with PEDS HEM/ONC or PICU for admission. Perform further evaluation and management as clinically appropriate (e.g., fluid resuscitation, vancomycin, etc.) → ADMIT TO PICU or PEDS HEM/ONC
  - Yes → Administer IV ceftriaxone + oral azithromycin → ADMIT TO PICU or PEDS HEM/ONC (based on supplemental O_2 need)
- No → Administer IV ceftriaxone → Age ≥ 18 months?
  - No → ADMIT TO PEDS HEM/ONC
  - Yes → Low risk?***
    - No → ADMIT TO PEDS HEM/ONC
    - Yes → Observe for 1 hour post antibiotics → Contact PEDS HEM/ONC to discuss discharge, follow-up, and additional antibiotic therapy

Antibiotics	Dose
Ceftriaxone IV	50 mg/kg/dose (max 2 g)
Azithromycin PO	10 mg/kg (max 500 mg/dose)

****Acute Chest Syndrome:**
New infiltrate on chest x-ray **AND** one or more of the following **NEW** symptoms:

- Fever
- Cough
- Sputum production
- Dyspnea
- Hypoxia

Fluids in Acute Chest Syndrome:
Avoid excessive fluid administration, resuscitate as necessary

*****Low Risk Features:**

- Normal vital signs
- Tolerating PO
- Hgb > 6 mg/dL
- Reticulocyte count > 5% (unless Hgb > 10 mg/dL)
- No significant drop in Hgb from baseline
- WBC 5,000-30,000/mm^3
- No history of bacteremia or sepsis
- No splenic sequestration within the past 3 months
- Not multiple visits for same febrile illness
- Compliant with penicillin
- Fully immunized
- Able to follow-up next day with Peds Hem/Onc via appointment or telephone contact

Contact CCKM for revisions.
Reference the UW Health Acute Evaluation and Management of Children with Sickle Cell Disease – Pediatric – ED Guideline

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