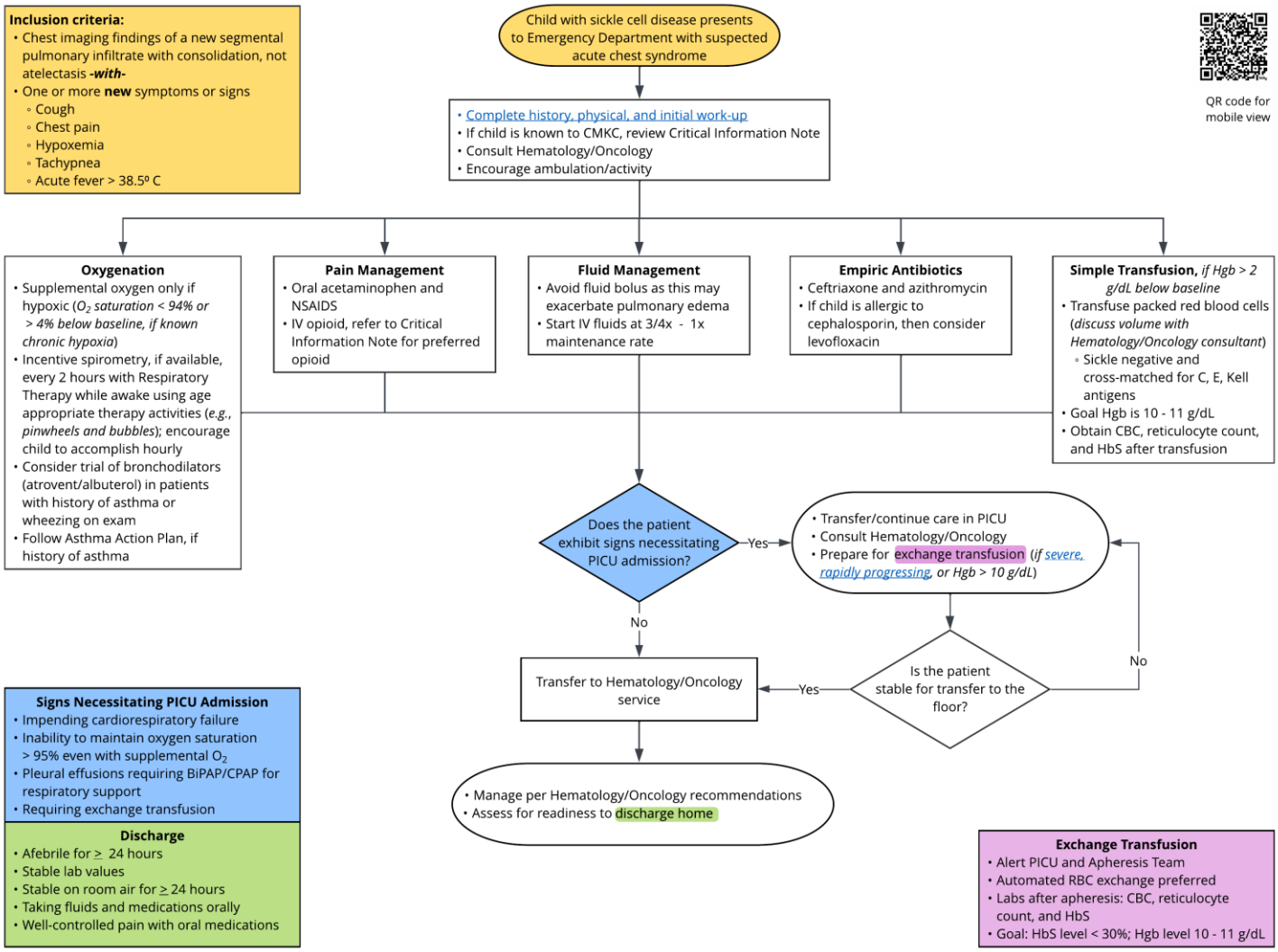
Sickle Cell Disease: Acute Chest Syndrome
Clinical Pathway Synopsis

Sickle Cell Disease: Acute Chest Syndrome Algorithm



** These clinical pathways do not establish a standard of care to be followed in every case. It is recognized that each case is different, and those individuals involved in providing health care are expected to use their judgment in determining what is in the best interests of the patient based on the circumstances existing at the time. It is impossible to anticipate all possible situations that may exist and to prepare a clinical pathway for each. Accordingly, these clinical pathways should guide care with the understanding that departures from them may be required at times.*

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Objective of Clinical Pathway

To provide care standards for the child, adolescent, or adult with sickle cell disease who presents with suspected acute chest syndrome. The Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway guides the early diagnosis and management of this potentially life-threatening complication of sickle cell disease when presenting to the emergency department.

Background/Epidemiology

Acute chest syndrome (ACS) is characterized by a new pulmonary infiltrate on chest radiographs accompanied by signs or symptoms that can include fever, cough, chest pain, respiratory distress, and hypoxia (Centers for Disease Control and Prevention, 2024; Howard et al., 2015; Jain et al., 2017; National Heart, Lung, and Blood Institute, 2014). The development of ACS is commonly associated with vaso-occlusive pain crisis in children with sickle cell disease, often necessitating hospitalization (Centers for Disease Control and Prevention, 2024; Howard et al., 2015; Jain et al., 2017; National Heart, Lung, and Blood Institute, 2014). Risk factors that typically predispose and increase the likelihood of developing ACS include infections, asthma, dehydration, or history of a recent surgical procedure (Centers for Disease Control and Prevention, 2024; Howard et al., 2015; Jain et al., 2017; National Heart, Lung, and Blood Institute, 2014).

Early diagnosis and management are essential to preventing progression to respiratory failure, multiorgan failure, or premature mortality (Centers for Disease Control and Prevention, 2024; Howard et al., 2015; Jain et al., 2017; National Heart, Lung, and Blood Institute, 2014). The Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway Committee aims to guide providers through the clinical detection of ACS and supportive care that includes oxygenation, pain management, fluid management, empiric antibiotic therapy, and transfusions to manage the child, adolescent, or adult with sickle cell disease presenting with suspected acute chest syndrome.

Target Users

- Physicians (Emergency Medicine, Hospital Medicine, Intensivists, Hematology/Oncology, Fellows, Residents)
- Advanced Practice Providers
- Nurses
- Respiratory Therapists

Target Population

Inclusion Criteria

- A child with sickle cell disease presenting with suspected acute chest syndrome:
  - Chest imaging findings of a new segmental pulmonary infiltrate with consolidation, not atelectasis
    – with-
  - One or more **new** symptoms or signs: cough, chest pain, hypoxemia, tachypnea, acute fever > 38.5° C

AGREE II

Two national guidelines (Chou et al., 2020; National Heart, Lung, and Blood Institute, 2014) and one international guideline (Howard et al., 2015) provided guidance to the Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway Committee. See Table 1, Table 2, and Table 3 for AGREE II.

Table 1

AGREE II Summary for the American Society of Hematology 2020 Guidelines for Sickle Cell Disease: Transfusion Support (Chou et al., 2020)

Domain	Percent Agreement	Percent Justification^
Scope and purpose	100%	The aim of the guideline, the clinical questions posed, and the target populations **were** identified.
Stakeholder involvement	92%	The guideline **was developed** by the appropriate stakeholders and represents the views of its intended users.

Rigor of development	91%	The process used to gather and synthesize the evidence, the methods to formulate the recommendations, and to update the guidelines **were** explicitly stated.
Clarity and presentation	100%	The guideline recommendations **are** clear, unambiguous, and easily identified; in addition, different management options are presented.
Applicability	89%	Barriers and facilitators to implementation, strategies to improve utilization, and resource implications **were addressed** in the guideline.
Editorial independence	100%	The recommendations **were not** biased by competing interests.
Overall guideline assessment	95%	
See Practice Recommendations		

Note: Four EBP Scholars completed the AGREE II on this guideline.
^ Percentage justification is an interpretation based on the Children's Mercy EBP Department standards.

Table 2
AGREE II Summary for the Evidence-Based Management of Sickle Cell Disease: Expert Panel Report (National Heart, Lung, and Blood Institute, 2014)

Domain	Percent Agreement	Percent Justification^
Scope and purpose	84%	The aim of the guideline, the clinical questions posed, and the target populations **were** identified.
Stakeholder involvement	92%	The guideline **was developed** by the appropriate stakeholders and represents the views of its intended users.
Rigor of development	93%	The process used to gather and synthesize the evidence, and the methods to formulate the recommendations **were** explicitly stated.
Clarity and presentation	100%	The guideline recommendations **are** clear, unambiguous, and easily identified; in addition, different management options are presented.
Applicability	82%	Barriers and facilitators to implementation, strategies to improve utilization, and resource implications **were addressed** in the guideline.
Editorial independence	83%	The recommendations **were not** biased by competing interests.
Overall guideline assessment	89%	
See Practice Recommendations		

Note: Four EBP Scholars completed the AGREE II on this guideline.
^ Percentage justification is an interpretation based on the Children's Mercy EBP Department standards.

Table 3
AGREE II Summary for the Guideline on the Management of Acute Chest Syndrome in Sickle Cell Disease (Howard et al., 2015)

Domain	Percent Agreement	Percent Justification^
Scope and purpose	90%	The aim of the guideline, the clinical questions posed, and the target populations **were** identified.
Stakeholder involvement	88%	The guideline **was developed** by the appropriate stakeholders and represents the views of its intended users.
Rigor of development	70%	The process used to synthesize the evidence and the methods to update the guidelines **were** explicitly stated. The guideline developers **did not** provide how the evidence was gathered or how the recommendations were formulated.

Clarity and presentation	97%	The guideline recommendations **are** clear, unambiguous, and easily identified. Different management options are also presented.
Applicability	48%	The guideline **addressed** barriers and facilitators to implementation. However, it **did not** include utilization strategies or resource costs associated with implementation.
Editorial independence	52%	It is **unclear** if the recommendations were biased by competing interests.
Overall guideline assessment	74%	
See Practice Recommendations		

Note: Four EBP Scholars completed the AGREE II on this guideline.
^ Percentage justification is an interpretation based on the Children's Mercy EBP Department standards.

Practice Recommendations

Please refer to the Guideline on the Management of Acute Chest Syndrome in Sickle Cell (Howard et al., 2015), the Evidence-Based Management of Sickle Cell Disease: Expert Panel Report (National Heart, Lung, and Blood Institute, 2014) and the American Society of Hematology guidelines (Chou et al., 2020) for evaluation and treatment recommendations.

Additional Questions Posed by the Clinical Pathway Committee

No clinical questions were posed for this review.

Updates from Previous Versions of the Clinical Pathway

- Oxygenation
  - Incentive spirometry continues to be encouraged. However, the spirometers are no longer supplied in the Emergency Department. Therefore, the recommendation for incentive spirometry is based on availability
  - The recommendation to complete intermittent positive pressure breathing every four hours when indicated has been removed, as it is no longer occurring in the Emergency Department
- Fluid Management
  - The recommendation for "nothing by mouth" was removed
- Empiric Antibiotic Therapy
  - Levofloxacin has replaced clindamycin as the alternative antibiotic to consider if the child has a cephalosporin allergy
- Signs Necessitating PICU Admission
  - Impending cardiorespiratory failure has replaced multilobar disease without pleural effusion

Recommendation Specific for Children's Mercy

There were no deviations from the Guideline on the Management of Acute Chest Syndrome in Sickle Cell (Howard et al., 2015), the Evidence-Based Management of Sickle Cell Disease: Expert Panel Report (National Heart, Lung, and Blood Institute, 2014), and the American Society of Hematology guidelines (Chou et al., 2020) regarding practice recommendations. However, logistical processes specific to Children's Mercy Kansas City were added.

- Clinical signs necessitating admission to the Pediatric Intensive Care Unit were provided
- Considerations regarding preparation for an exchange transfusion were included
- Discharge criteria were provided

Measures

- Use of Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway
- Use of the associated powerplan(s)

Value Implications

The following improvements may increase value by reducing healthcare costs and non-monetary costs (e.g., missed school/work, loss of wages, stress) for patients and families and reducing costs and resource utilization for healthcare facilities.

- Decreased risk of delayed recognition and management of acute chest syndrome in children with sickle cell disease
- Decreased inpatient length of stay
- Decreased unwarranted variation in care

Organizational Barriers and Facilitators

Potential Barriers

- Variability of an acceptable level of risk among providers
- Challenges with follow-up faced by some families

Potential Facilitators

- Collaborative engagement across care continuum settings during clinical pathway development
- High rate of use of the clinical pathway
- Standardized order set for Emergency Department, Hospital Medicine, and Pediatric Intensive Care

Bias Awareness

Bias awareness is our aim to recognize social determinants of health and minimize healthcare disparities, acknowledging that our unconscious biases can contribute to these inequities.

Power Plans

- EDP Sickle Cell with Chest Pain Pathway
- EDP Sickle Cell with Fever ED Standing Orders
- EDP Sickle Cell Continuous PCA Infusion
- Sickle Cell Simple Transfusion
- Sickle Cell Acute Chest Syndrome Admission

Associated Policies

- Sickle Cell Disease with Fever Standing Order
- Sickle Cell Related Pain
- Sickle Cell Disease with Pain Standing Order

Education Materials

- Acute Chest Syndrome
    - Found in Cerner depart process
    - Available in English and Spanish

Clinical Pathway Preparation

This pathway was prepared by the Evidence Based Practice (EBP) Department in collaboration with the Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway Committee composed of content experts at Children's Mercy Kansas City. If a conflict of interest is identified, the conflict will be disclosed next to the committee member's name.

Clinical Pathway Representation

This clinical pathway was originally created with representation from Hematology/Oncology/BMT Division, Critical Care Medicine, Pediatric Emergency Medicine, Nursing, Respiratory Therapy, Medical Informatics, and Evidence-Based Practice.

Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway Committee Members and Representation

- Shabnam Arsiwala, MD, FAAP | Hematology/Oncology/BMT | Committee Co–Chair
- Vivek Dubey, MD | Pediatric Emergency Medicine | Committee Co-Chair

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- Jay Rilinger, MD | Critical Care Medicine | Committee Member
- Megan Gubichuk, MD | Pulmonology | Committee Member
- Sarah Dierking, MSN, RN, CPHQ | Clinical Practice and Quality | Committee Member

EBP Committee Members

- Todd Glenski, MD, MSHA, FASA | Anesthesiology, Evidence Based Practice
- Kelli Ott, OTD, OTR/L | Evidence Based Practice

Clinical Pathway Development Funding

The development of this clinical pathway was underwritten by the following departments/divisions: Pediatric Emergency Medicine, Hematology/Oncology/BMT, Critical Care Medicine, Pulmonology, Clinical Practice and Quality, and Evidence Based Practice

Conflict of Interest

The contributors to the Sickle Cell Disease: Acute Chest Syndrome Clinical Pathway have no conflicts of interest to disclose related to the subject matter or materials discussed.

Approval Process

- This pathway was reviewed and approved by the EBP Department and the Sickle Cell Disease: Acute Chest Syndrome Committee after committee members garnered feedback from their respective divisions/departments.

Review Requested

Department/Unit	Date Obtained
Hematology/Oncology/BMT	June 2025
Pediatric Emergency Medicine	June 2025
Critical Care Medicine	June 2025
Pulmonology	July 2025
Clinical Practice and Quality	July 2025
Evidence Based Practice	June 2025

Version History

Date	Comments
January 2021	Version one – (*algorithm and associated powerplan developed*)
July 2025	Version two – (*algorithm revised, associated powerplans reviewed, and associated synopsis developed*)

Date for Next Review

- July 2028

Implementation & Follow-Up

- Once approved, the pathway was implemented and presented to appropriate care teams:
  - Announcements were made to the relevant departments
- Order sets consistent with recommendations were updated
- The policies were reviewed. This details a process for nursing staff to improve comfort and expedite care for children with sickle cell disease who present with fever or experience a painful episode
- Care measurements may be assessed and shared with appropriate care teams to determine if changes need to occur
- Pathways are reviewed every 3 years (or sooner) and updated as necessary within the EBP Department at CMKC. Pathway committees are involved with every review and update

Disclaimer

When evidence is lacking or inconclusive, options in care are provided in the supporting documents and the power plan(s) that accompany the clinical pathway.

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