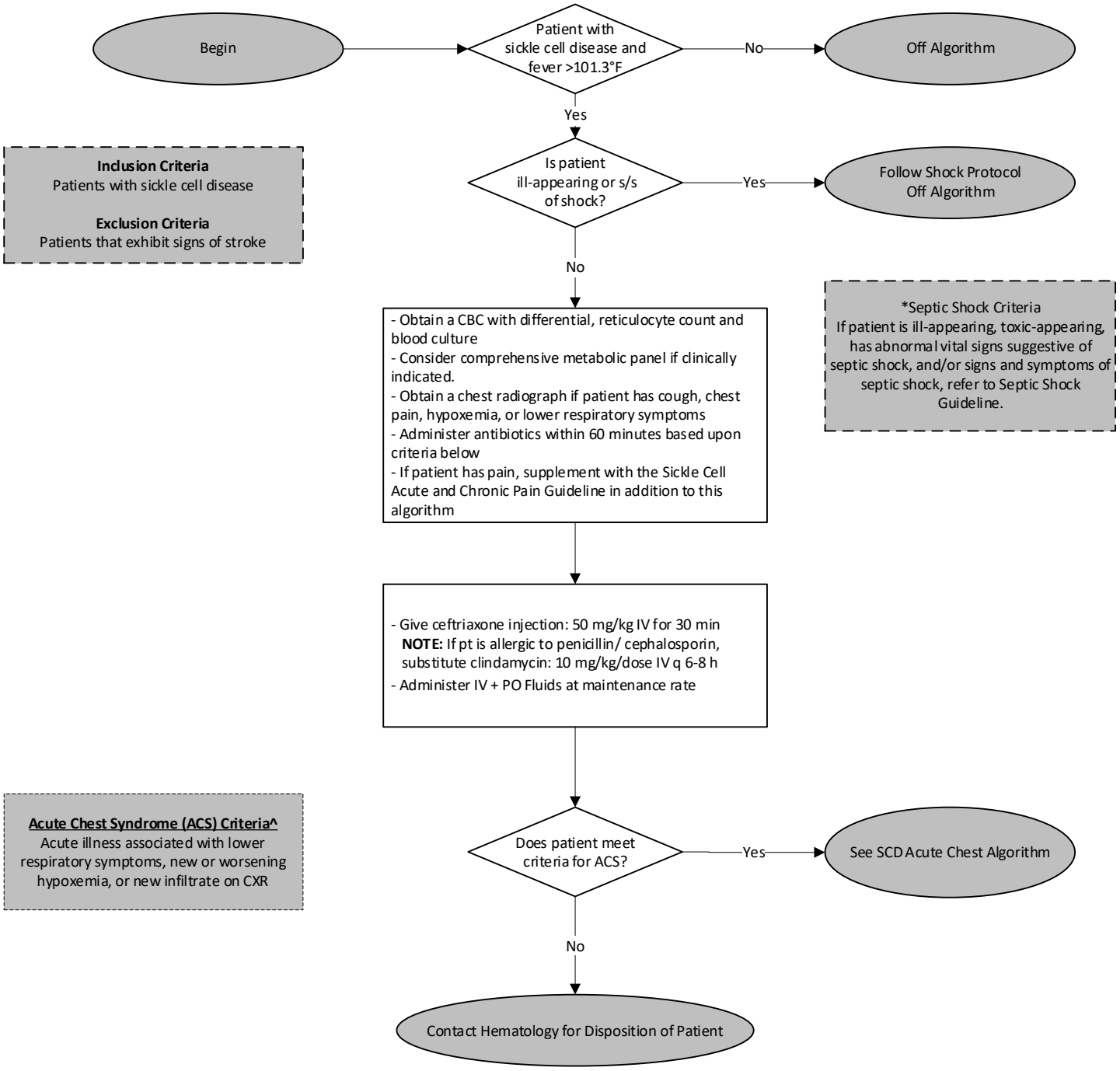
TEXAS CHILDREN'S HOSPITAL
EVIDENCE-BASED OUTCOMES CENTER
Sickle Cell Disease With Fever
Evidence-Informed Pathway



Critical Points of Evidence*

Evidence Supports

- Obtain a chest radiograph if patient has cough, chest pain, hypoxemia, or lower respiratory symptoms at presentation to the hospital or during admission. Fever in the absence of these symptoms does not necessitate evaluation with a chest radiograph. [1-6] - Strong recommendation with very low quality evidence
- Encourage oral fluid intake and to administer total fluid intake (intravenous plus oral) at maintenance rate to encourage that the patient remains euvolemic. [7-9] - Strong recommendation with very low quality evidence
  Remarks: Utilize ¾ maintenance fluid if the patient is suspected or confirmed to have acute chest syndrome.
- Use continuous pulse oximetry monitoring in patients with sickle cell disease with fever who are being treated in the emergency center. [10-17] - Strong recommendation with very low quality evidence

Recommendations Adopted/Adapted from National Guidelines

- Obtain a CBC with differential, reticulocyte count and blood culture for all children with fever and sickle cell disease. Consider comprehensive metabolic panel if clinically indicated. [18]
  Remarks: Hemoglobin and retic count should be interpreted relative to baseline values
  - Anemia + low retic = concern for aplastic crisis
  - Anemia + low platelet count + normal retic = concern for splenic sequestration
    Anemia can progress extremely quickly due to low baseline hemoglobin and short half-life of RBCs due to clearance of abnormal cells. This recommendation was adapted from the American College of Emergency Physicians Guideline.
- Promptly administer empiric ceftriaxone for children with SCD and fever. [6]
  Remarks: This recommendation was adapted from the National Institutes of Health Guideline.

*NOTE: The references cited represent the entire body of evidence reviewed to make each recommendation.

Measures

Process

- Frequency of ambulation

Outcome

- Number of patients who develop ACS
- Number of patients transferred to critical care
- ED and IP LOS

References

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Clinical Standards Preparation

This clinical standard was prepared by the Evidence-Based Outcomes Center (EBOC) team in collaboration with content experts at Texas Children's Hospital. Development of this clinical standard supports the TCH Quality and Patient Safety Program initiative to promote clinical standards and outcomes that build a culture of quality and safety within the organization.

Sickle Cell Disease With Fever and Acute Chest Syndrome Content Expert Team

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No relevant financial or intellectual conflicts to report.

Development Process

This clinical standard was developed using the process outlined in the EBOC Manual. The literature appraisal documents the following steps:

1. Review Preparation
   - PICO questions established
   - Evidence search confirmed with content experts
2. Review of Existing External Guidelines
   - National Institute of Health, Evidence-Based Management of Sickle Cell Disease Expert Panel Report, 2014; National Institute for Health Care Excellence (NICE), Sickle Cell Disease: Managing Acute Painful Episodes in Hospital, 2012; American College of Emergency Physicians, Sickle Cell – Point of Care Tools, 2023;
3. Literature Review of Relevant Evidence
   - Searched: PubMed, Cochrane Collaboration
4. Critically Analyze the Evidence
   - Fifteen nonrandomized studies
5. Summarize the Evidence
   - Materials used in the development of the clinical standard, literature appraisal, and any order sets are maintained in an electronic database.

Evaluating the Quality of the Evidence

Published clinical guidelines were evaluated for this review using the **AGREE II** criteria. The summary of these guidelines are included in the literature appraisal. AGREE II criteria evaluate Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity and Presentation, Applicability, and Editorial Independence using a 4-point Likert scale. The higher the score, the more comprehensive the guideline.

This clinical standard specifically summarizes the evidence *in support of* or *against* specific interventions and identifies where evidence is *lacking/inconclusive*. The following categories describe how research findings provide support for treatment interventions.
"Evidence Supports" provides evidence to support an intervention
"Evidence Against" provides evidence against an intervention.
"Evidence Lacking/Inconclusive" indicates there is insufficient evidence to support or refute an intervention and no conclusion can be drawn *from the evidence.*
The **GRADE** criteria were utilized to evaluate the body of evidence used to make practice recommendations. The table below defines how the quality of the evidence is rated and how a strong versus weak recommendation is established. The literature appraisal reflects the critical points of evidence.

Recommendation	
STRONG	Desirable effects clearly outweigh undesirable effects or vice versa
WEAK	Desirable effects closely balanced with undesirable effects
Quality	**Type of Evidence**
High	Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies
Moderate	Evidence from RCTs with important limitations (e.g., inconsistent results, methodological flaws, indirect evidence, or imprecise results) or unusually strong evidence from unbiased observational studies
Low	Evidence for at least 1 critical outcome from observational studies, RCTs with serious flaws or indirect evidence
Very Low	Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence

Recommendations

Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible. The Content Expert Team and EBOC team remain aware of the controversies in the diagnosis/management of sickle cell disease with fever and acute chest syndrome in children. When evidence is lacking, options in care are provided in the clinical standard and the accompanying order sets (if applicable).

Approval Process

Clinical standards are reviewed and approved by hospital committees as deemed appropriate for its intended use. Clinical standards are reviewed as necessary within EBOC at Texas Children's Hospital. Content Expert Teams are involved with every review and update.

Disclaimer

Practice recommendations are based upon the evidence available at the time the clinical standard was developed. Clinical standards (guidelines, summaries, or pathways) do not set out the standard of care and are not intended to be used to dictate a course of care. Each physician/practitioner must use his or her independent judgment in the management of any specific patient and is responsible, in consultation with the patient and/or the patient's family, to make the ultimate judgment regarding care.

Version History

Date	Comments
2/2024	Originally Completed