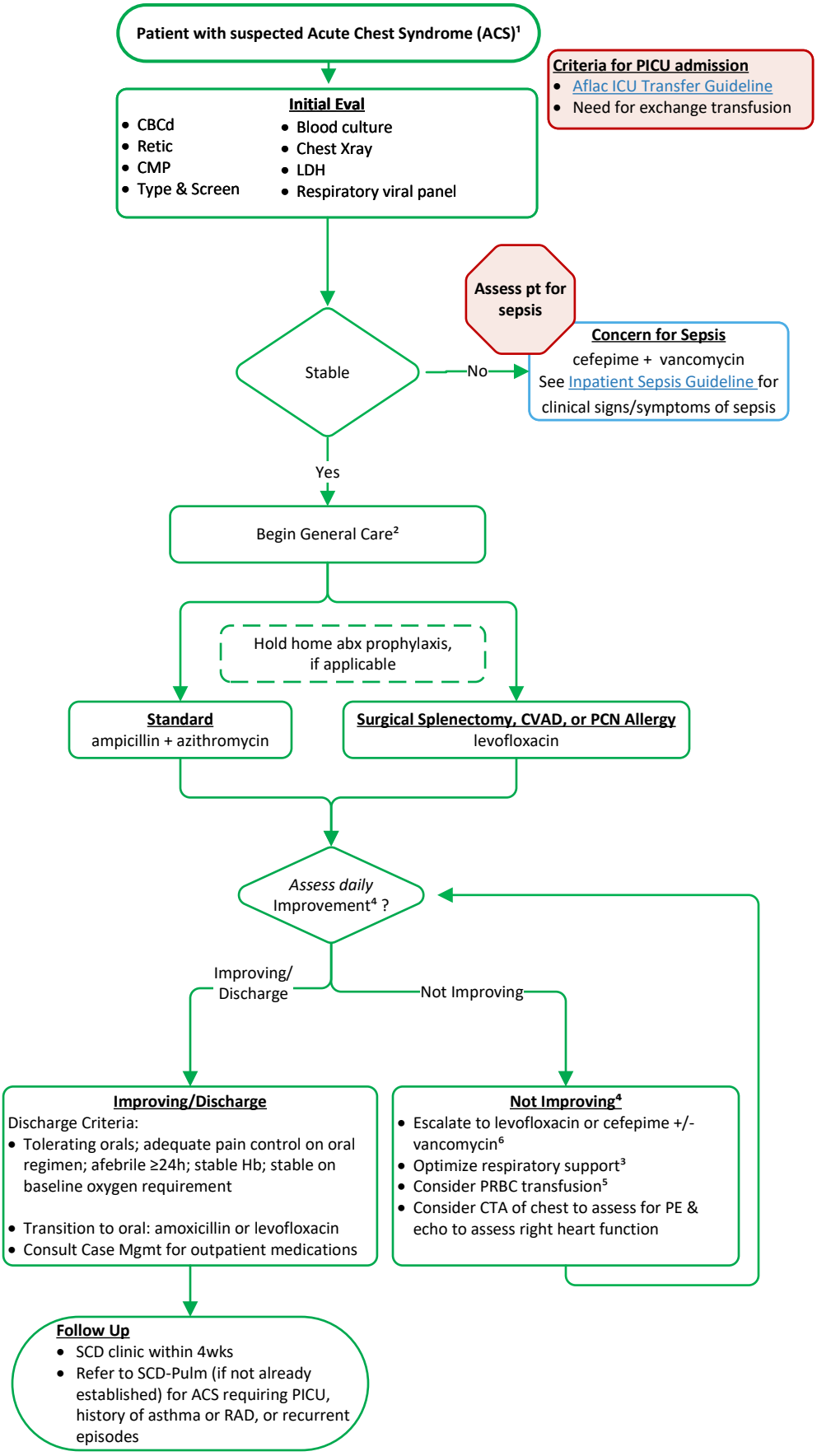
Sickle Cell Disease (SCD) Acute Chest Syndrome (ACS) Pathway
IP Management

December 2025

Children's Healthcare of Atlanta

Work Up



[1] Definition

Acute illness characterized by new pulmonary infiltrate (non-atelactatic consolidation) on CXR plus fever (≥ 38.3C) and/or respiratory signs/symptoms (cough, shortness of breath, chest pain, crackles, hypoxia, etc)

[2] General Care

- Maintain euvolemia with IV fluids at 3/4 to 1x maintenance rate. Decrease IVF rate as clinical status improves and oral intake increases.
- Consider PT consult for early ambulation
- Consider VTE prophylaxis, especially in patients >18yo, previous history of VTE, CVL, or other risk factors
- See SCD Inpatient Pain Guideline

Daily Labs

- CBCd
- Retic
- Ensure active Type & Screen

[3] Respiratory Support

- Keep oxygen saturations ≥ 93%
- All patients should receive positive expiratory pressure (PEP) and incentive spirometer (IS)

Consider the following:

- Scheduled albuterol if history of asthma, reactive airway disease, or history of severe ACS
- 3% inhaled saline and/or chest physiotherapy (CPT) for patients with significant crackles/mucous
- Pulmonary consult based on severity, history of recurrence, or other lung disease
- Positive pressure ventilation (PPV) such as HFNC or BiPAP for increasing WOB, O2 needs, or previous recommendations to start PPV at dx of ACS

[4] Non Improvement Criteria

If:

- Increasing respiratory support
- Worsening imaging
- Hemodynamic instability or concern for sepsis

Then:

- Consider Watcher Huddle

[5] Transfusion

- RBC Transfusion has benefits to treat ACS with hypoxia. Balance benefits vs individual risk assessment for each patient
- Assess Hb, retic, HbS%, alloimmunization history, level of respiratory support

If decision is made to transfuse and:

- Hb < 9, can proceed with simple transfusion
- Hb ≥ 9 and/or rapid deterioration characterized by increasing oxygen needs, worsening respiratory distress, progressive pulmonary infiltrates, and/or decline in Hb despite simple transfusions, consider exchange transfusion with a target HbS% <30%

[6] Consider vancomycin if...

- Moderate to severe parapneumonic effusion
- Empyema
- Other concern for MRSA (obtain nasal MRSA PCR)

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Medication Table				
Medication	**Route**	**Dose**	**Frequency**	**Notes**
ampicillin	IV	50 mg/kg *Max 2000 mg/dose*	Q6h	
cefepime	IV	50 mg/kg *Max 2000 mg/dose*	Q8h	
vancomycin	IV	20 mg/kg *Max 1000 mg/dose*	Q8h	Need to check levels if remaining on vancomycin after 48h
azithromycin	IV/PO	10 mg/kg *Max 500 mg/dose*	Daily x 3 days	Total course 3 days
levofloxacin	IV/PO	10 mg/kg *Max 750 mg/day*	Q12h for < 5 y.o Q24h for >/= 5 y.o.	Total course IV+PO = 7-10 days based on individual patient
amoxicillin	IV/PO	90 mg/kg/day divided *Max 1000 mg/dose*	BID	Total course IV+PO = 7-10 days based on individual patient

References

1. National Heart, Lung, and Blood Institute. Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014.
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3. Gillespie M, Afolabi-Brown O, Machogu E, Willen S, Kopp BT. Updates in Pediatric Sickle Cell Lung Disease. Clin Chest Med. 2024 Sep;45(3):749-760. doi:10.1016/j.ccm.2024.02.022. PMID: 39069335.
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5. Mekontso Dessap, A., Dauger, S., Khellaf, M. et al. Guidelines for the management of emergencies and critical illness in pediatric and adult patients with sickle cell disease. Ann. Intensive Care 15, 74 (2025). https://doi.org/10.1186/s13613-025-01479-3

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