Inclusion Criteria
All sickle cell patients in the ED or inpatient units admitted for any reason

Perform Incentive Spirometry (I.S.)
- 10 breaths
- Prior to chest X-rays
- May use developmentally appropriate alternative as needed (blowing bubbles, pinwheel, etc.)

ED
- q2 hrs while awake

Inpatient
- q2 hrs from 0800 to 2200
- With vital signs while awake from 2200 to 0800
- With every "as needed" IV bolus of pain

Pain Management Considerations
- Assure pain is vaso-occlusive
- Evaluate dose
- Assess efficacy of pain management plan
- Evaluate for use of adjuvants such as muscle relaxants, benzodiazepines, ketamine
- Consider regional and spinal blocks
- Consider topical therapies
- Consider Bi-PAP or C-PAP
- Ensure non-pharmacological methods are in place such as heat massage, acupuncture, distraction, etc.

Provide Feedback
- Positive reinforcement
- Remind why important

Assess Barriers & Intervene

Too Much Pain?
- Offer PRN dose
- Reassess pain & ability to perform I.S. 15-20 min after PRN dose
- If pain not improved after 3 boluses (in approximately 30 min), re-evaluate and adjust pain plan with team
- See pain management considerations

Too Sedated?
- Evaluate sedation level
- HOLD their next IV bolus or short acting opiate and have the medical team re-evaluate
- See pain management considerations

Behavioral Reasons?
- Patient & family coaching
- I.S. is necessary to prevent life threatening respiratory complications
- Opiates cannot be given safely unless I.S. is done
- Inform that PRN doses will not be given for 4 hr if I.S. not done
- Family support – contact guardian if not available
- Follow patient specific behavior plan if available
- Hold next opiate dose if I.S. not done and inform that opiates may need to be weened

Go to Transfusion Pathway

For questions concerning this pathway, contact: SickleCellPathway@seattlechildrens.org
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Last Updated: February 2019
Next Expected Review: July 2019
**Inclusion Criteria**
- New, non-atelactatic, pulmonary infiltrate in a patient with a sickle hemoglobinopathy

**Exclusion Criteria**
- O₂ need related to opiate use with no infiltrate on CXR

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**Evaluations for Acute Chest**
- Physical Exam
- O₂ sats
- Blood & Urine Cx
- CXR
- Consider viral studies

**Standard Conservative Therapy**
- Aggressive pain management
- Incentive spirometry q 1 hrs while awake, 10 breaths every hour from 0800 to 2200 and with vital signs while awake from 2200 to 0800, as well as with every "as needed" IV bolus of pain medication, and prior to chest x-rays
- Oxygen to maintain O₂ saturation >93%
- Ceftriaxone q 24 hours IV (Ciprofloxacin and Clindamycin if allergic to Ceftriaxone)
- Azithromycin or other macrolide antibiotic to cover atypical organisms
- Consider bronchodilators
- Patients with a consolidation on CXR should be evaluated by respiratory therapy for CPT
- Consider CPAP or BiPAP for patients with poor respiratory effort, reduced ventilation or continued decline despite other interventions

**Assess for Immediate Transfusion**
- Critically ill
- Multi-lobe Involvement
- PaO₂ <70 or 10mm below baseline
- O₂ by facemask requirement with HCT <20%

**Assess for Progressive Pulmonary Decline**
- Persistent increase in O₂ need (not a transient need) of >1L/shift or 2L/ 24 hours
- Steady worsening of physical exam not attributable to fluid overload
- Increasingly toxic appearance
- Substantially worsened CXR, acknowledging CXR changes often lag behind clinical status

**Hematocrit Evaluation**
- HCT <18%
- See Transfusion Guidelines for detailed transfusion criteria for anemia and other sickle cell transfusion indications

**Transfuse Immediately Targeting HCT of 30-33%**
- If Hct <27%
  - Consider direct transfusion
- If Hct ≥27%
  - Consider exchange transfusion

**Attributes**
- HbS neg, leukoreduced, antigen matched

See Transfusion Guidelines for details on how much blood to transfuse

**Discharge Criteria**
- Improved respiratory symptoms, off O₂ and O₂ sats at baseline.
- Afebrile for 24 hrs and negative cultures for 24 hrs if applicable.
- Adequate oral intake, and able to take oral antibiotics.
- Adequate pain relief (if needed) with oral analgesics and written taper of opiates given to patient and reviewed.
- Discharge teaching on home use of incentive spirometry (if also taking opiates) completed.
- Stable Hct and reticulocyte count.
- Follow-up plans coordinated with sickle cell service
- CXR 8-12 weeks post therapy
- Consider follow up pulmonary function tests

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Incentive Spirometry

- Perform incentive spirometry, 10 breaths q 2 hours while awake from 0800 to 2200 and with vital signs while awake from 2200 to 0800, as well as with every “as needed” IV bolus of pain medication, and prior to chest x-rays

[LOE: ★★★★ Very Low Quality] (Bellet, 1995)
• If critically ill, multi lobe involvement or a PaO$_2$ <70 or 10mm below baseline transfuse immediately targeting a Hct of 30-33%. Regardless of ACS consider transfusion for a Hct of <18% per standard transfusion for anemia guidelines. Similarly, consider transfusion for patients with Hct <20% who require oxygen by facemask (i.e. not on facemask for comfort).

• For progressive pulmonary decline despite other interventions (especially if Hct>5 % points below baseline or Hct under 18%) consider a direct transfusion targeting a Hct of 30-33% (do not transfuse acutely to Hct>33%). Progressive decline could include:
  • persistent increase in O2 need (not a transient need) of >1L/ shift or 2L/ 24 hours.
  • steady worsening of physical exam not attributable to fluid overload
  • increasingly toxic appearing
  • mental status changes not attributable to opiates
  • substantially worsened CXR, acknowledging CXR changes often lag behind clinical status

[LOE: (E) Expert Opinion] (Vichinsky, 2000; Wun, 2009; Miller, 2002)
Assess Sedation Level

- Attempt to elicit patient response (utilize parent or caregiver if available)
  - Verbal communication
    - State your name, patient name, and that it's time to do incentive spirometry
    - If asleep, ask patient to open eyes or sit up
  - Gentle tactile stimulation to non-painful area
    - Touch arm, rub palm, etc. after assuring this is not an area where patient has sickle cell pain.
  - Gentle shaking to non-painful area
  - Continue to repeat the above continuously for 1-2 minutes
  - If unable to elicit response, remind the patient of the concern of over sedation and the risk of developing acute chest syndrome and the consequence of eliminating PRN pain medications for 4 hours if I.S. is not performed.

- After any response, re-attempt Incentive Spirometry

- If unable to elicit a response, the patient is too sedated to receive a PRN bolus
  - Remind patient of concerns of over sedation and the risk of acute chest syndrome and consequence of eliminating PRN pain medications for 4 hours if I.S. is not performed.
  - Notify team for need to re-evaluate of pain plan

[LOE: (E) Local Consensus]
We used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial, or observational studies. The rating is then adjusted in the following manner:

Quality ratings are downgraded if studies:
• Have serious limitations
• Have inconsistent results
• If evidence does not directly address clinical questions
• If estimates are imprecise OR
• If it is felt that there is substantial publication bias

Quality ratings can be upgraded if it is felt that:
• The effect size is large
• If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
• If a dose-response gradient is evident

**Quality of Evidence:**
- ★★★★★ High quality
- ★★★★ Moderate quality
- ★★★ Low quality
- ★★★☆ Very low quality
- ★☆☆☆ Expert Opinion (E)

Title: Sickle Cell Pathway

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Date: June 2013

Retrieval Website: http://www.seattlechildrens.org/pdf/Sickle-Cell-Algorithm.pdf

Example:
Summary of Version Changes

- **Version 1.1 (4/15/2013)**: Go live
- **Version 1.2 (3/20/2015)**: Algorithm wording correction to align with powerplan.
- **Version 1.3 (02/08/2019)**: Contact e-mail updated
Medical Disclaimer

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children’s Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

Readers should confirm the information contained herein with other sources and are encouraged to consult with their health care provider before making any health care decision.
Studies were identified by searching electronic databases using search strategies developed and executed by a medical librarian, Susan Klawansky. Searches were performed in June 2011 in the following databases: on the Ovid platform – Medline (1970 to current), Medline in Process, Cochrane Database of Systematic Reviews (2005 to current), Cochrane Central Register of Controlled Trials (all), Health Technology Assessment (all); elsewhere – CINAHL (all), Clinical Evidence, DynaMed, UpToDate, AHRQ, National Guideline Clearinghouse, American Academy of Pediatrics Policies, ClinicalTrials.gov and TRIP. Retrieval was limited to humans of any age and no language or other restrictions were imposed. In Medline, appropriate Medical Subject Headings (MeSH) were used, along with text words, and the search strategy was adapted for other databases using their controlled vocabularies, where available, along with text words. Concepts searched were sickle cell anemia, acute chest syndrome, thalassemia and incentive spirometry or positive expiratory pressure.

Susan Klawansky, MLS, AHIP
June 29, 2012

Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535
Literature Search

Studies were identified by searching electronic databases using search strategies developed and executed by a medical librarian, Susan Klawansky. Searches were performed in June 2011 in the following databases: on the Ovid platform – Medline (1980 to current), Medline in Process, Cochrane Database of Systematic Reviews (2005 to current), Cochrane Central Register of Controlled Trials (all); elsewhere – Clinical Evidence, DynaMed, UpToDate, AHRQ, National Guideline Clearinghouse, American Academy of Pediatrics Policies, TRIP and Cincinnati Children’s Evidence-Based Guidelines. Retrieval was limited to humans of any age and English language. In Medline, appropriate Medical Subject Headings (MeSH) were used, along with text words, and the search strategy was adapted for other databases using their controlled vocabularies, where available, along with text words. Concepts searched were sickle cell anemia, thalassemia AND acute chest syndrome AND blood transfusion. All retrieval was further limited to certain evidence categories, such as relevant publication types, Clinical Queries, index terms for study types and other similar limits.

Susan Klawansky, MLS, AHIP
June 29, 2012

Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535
Incentive Spirometry


Transfusions