Incentive Spirometry in Sickle Cell Disease v1.2

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

Provide Feedback
- Positive reinforcement
- Remind why important

Patient performs I.S.
Patient does not perform I.S.

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

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Go to Transfusion Pathway

Re-attempt I.S.

Patient performs I.S.
Patient does not perform I.S.

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.
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

Transfusion for Acute Chest Syndrome in Sickle Cell v1.2

Executive Summary

Citation Information

Explanation of Evidence Ratings

Summary of Version Changes

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

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

Yes

No

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

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.

No

For questions concerning this pathway, contact: sicklecell@seattlechildrens.org

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Medical Disclaimer

Last Updated: March 2015
Next Expected Revision: April 2016
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)
Transfusion Guidelines for Acute Chest Syndrome

- If critically ill, multi lobe involvement or a PaO₂ <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
- ✷✷✷ O Moderate quality
- ✷✷ O Low quality
- ✷ O Very low quality
- Expert Opinion (E)

Executive Summary

Objective
To standardize the care of patients hospitalized with sickle cell disease, specifically the adherence to incentive spirometry guidelines and the appropriateness of transfusions in patients with Acute Chest Syndrome (ACS).

Recommendations
1. 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.
2. Transfuse patients with the following criteria:
   a. If critically ill, multi lobe involvement or a PaO2 <70 or 10mm below baseline transfuse immediately targeting a hematocrit of 30-33%. Regardless of ACS consider transfusion for a hematocrit of <18% per standard transfusion for anemia guidelines. Similarly, consider transfusion for patients with hematocrit <20% who require oxygen by facemask (i.e. not on facemask for comfort)
      OR
   b. For progressive pulmonary decline despite other interventions (especially if Hematocrit >5% points below baseline or Hematocrit under 18%) consider a direct transfusion targeting a Hematocrit of 30-33% (do not transfuse acutely to Hematocrit >33%).
      Progressive decline could include:
      i. Persistent increase in O2 need (not a transient need) of >1L/shift or 2L/ 24 hours.
      ii. Steady worsening of physical exam not attributable to fluid overload
      iii. Increasingly toxic appearing
      iv. Mental status changes not attributable to opiates
      v. Substantially worsened CXR, acknowledging CXR changes often lag behind clinical status

Rationale
- **Safety** will be improved by promoting adherence to incentive spirometry, which has been shown to be successful in decreasing the likelihood of developing acute chest syndrome in hospitalized sickle cell patients.
- **Quality** of care will be improved by reducing variability with standardization, incorporating the use of the best available evidence and consensus, and preventing the long term complications of iron overload and alloimmunization associated with transfusion as well as lung tissue damage associated with ACS.
- **Delivery** will be improved by aiming to only transfuse patients who meet specific criteria.
- **Engagement** is grounded in the fact that the pathway has been developed, reviewed, and vetted by all members of the sickle cell team and reviewed the Hematology / Oncology Practices and Procedures meeting.
- **Patient/Family Satisfaction** will be addressed by implementing clinical standard work that will assure the highest quality of care, and increasing consistency in care among members of the health care team.
Executive Summary

- Cost of care is anticipated to be more predictable and easier to measure and control with a more standardized process. A decreased incidence of ACS will decrease costs while improving quality of care. Reducing unnecessary transfusion will decrease costs and decrease the cost of managing transfusion related complications.

Evidence
A literature search was conducted by our librarian services in attempt to answer these clinical questions with the highest level of evidence. These references were further reviewed and their applicability to these questions summarized and documented to inform recommendations (please see the Evidence and Recommendations document for specific details).

Implementation Highlights
- Algorithm
- Recurring presentations at faculty and staff meetings
- CIS Ordersets

Metrics Plan
1. Core Clinical Effectiveness /CSW metrics:
   a. Count of Inpatient/observation discharges
   b. Median Length of Stay
   c. Percent of patients with any of the specified orderset
   d. Average charges per case
   e. Readmissions
2. Specific process metrics:
   a. Incentive spirometry nursing documentation compliance
   b. Incentive spirometry patient adherence
   c. The percent of sickle cell patients admitted for pain that develop ACS.
   d. Percent of patients with ACS who do not meet guidelines but receive transfusion
   e. Percent of patients with ACS who meet guidelines but do not receive transfusion

PDCA Plan
The CSW owner and committee will follow metrics, continue to review medical literature, and make alterations to the pathway as needed.

Revision History
Date Approved:       June 2013
Next Review Date:   June 2016
Executive Summary

**CSW Owner:**  
Bender and the Clinical Effectiveness Program

*Approved by the Sickle Cell Clinical Standard Work (CSW) Team in June 2013.*

**Sickle CSW Team:**
Owner: M. A. Bender, MD, PhD  
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Susan Klawansky, Librarian  
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- Michael Bender
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- Kate Drummond
- Jeff Foti
- Leah Kroon
- Ryan Leininger
- Michael Leu

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.
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
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Susan Klawansky, MLS, AHIP
June 29, 2012

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


Transfusions

- Thame JR; Hambleton IR; Serjeant GR: RBC transfusion in sickle cell anemia (HbSS): Experience from the Jamaican Cohort Study. Transfusion. 2001; 41(5): 596-601.