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Improving Care for Children With Sickle Cell Disease/Acute Chest Syndrome

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Pediatrics 2011;127;e480; originally published online January 17, 2011;
DOI: 10.1542/peds.2010-3099

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abstract

BACKGROUND: Acute chest syndrome (ACS) is a leading cause of hospitalization and death of children with sickle cell disease (SCD). An evidence-based ACS/SCD guideline was established to standardize care throughout the institution in February 2008. However, by the summer of 2009 use of the guideline was inconsistent, and did not seem to have an impact on length of stay. As a result, an implementation program was developed.

OBJECTIVE: This quality-improvement project evaluated the influence of the development and implementation of a clinical practice guideline for children with SCD with ACS or at risk for ACS on clinical outcomes.

METHODS: Clinical outcomes of 139 patients with SCD were evaluated before and after the development of the implementation program. Outcomes included average length of stay, number of exchange transfusions, average cost per SCD admission, and documentation of the clinical respiratory score and pulmonary interventions.

RESULTS: Average length of stay decreased from 5.8 days before implementation of the guideline to 4.1 days after implementation ($P = .033$). No patients required an exchange transfusion. Average cost per SCD admission decreased from $30 359 before guideline implementation to $22 368. Documentation of the clinical respiratory score increased from 31.0% before implementation to 75.5%, which is an improvement of 44.5% ($P < .001$). Documentation of incentive spirometry and positive expiratory pressure increased from 23.3% before implementation to 50.4%, which is an improvement of 27.1% ($P < .001$).

CONCLUSIONS: Implementation of a guideline for children with SCD with ACS or at risk for ACS improved outcomes for patients with SCD. *Pediatrics* 2011;127:e480–e488

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KEY WORDS

quality improvement, implementation, guideline development, acute chest syndrome, sickle cell disease

ABBREVIATIONS

ACS—acute chest syndrome
SCD—sickle cell disease
IS—incentive spirometry
PEP—positive expiratory pressure
CRS—clinical respiratory score
PDSA—plan-do-study-act

www.pediatrics.org/cgi/doi/10.1542/peds.2010-3099

doi:10.1542/peds.2010-3099

Accepted for publication Nov 29, 2010

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PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275).

FINANCIAL DISCLOSURE: *Dr Katkin serves on research advisory committees for 2 studies conducted by MedImmune, neither of which has any relationship to sickle cell disease or quality improvement; the other authors have indicated they have no financial relationships relevant to this article to disclose.*

Acute chest syndrome (ACS) usually presents in patients with sickle cell disease (SCD) as a rapidly occurring pulmonary disease recognized by lower respiratory tract symptoms, hypoxemia, and a new infiltrate shown on a chest radiograph.[1] ACS causes a high number of hospitalizations and deaths of children with SCD, and by some estimates, it accounts for as many as 25% of SCD-related deaths.[1–4]

Almost half of the patients diagnosed with ACS develop the respiratory complication during hospitalization for another diagnosis such as pain crisis. Factors such as young age, low baseline hemoglobin level, and low fetal hemoglobin level place patients at a higher risk of developing ACS.[3–7] Opioid analgesics, postoperative atelectasis, and asthma exacerbations or respiratory viral infections may promote the development of an ACS episode.[1–9] Although the etiology is undetermined in many cases, pulmonary infarction, fat embolism, and infection are now recognized as important causes of ACS. *Chlamydia pneumoniae*, *Mycoplasma pneumoniae*, *Streptococcus pneumoniae*, *Staphylococcus aureus*, and parvovirus are pathogens commonly isolated from patients with ACS.[2]

Management of ACS is primarily supportive care and includes respiratory therapy (eg, incentive spirometry [IS], positive expiratory pressure [PEP], and bronchodilators), antibiotics, and red-cell transfusions (J. Myers, unpublished PowerPoint presentation, 2006, and refs [10–22]). There have been few published studies proving the effectiveness of these measures in the management of ACS. In their study, Bellet et al[13] documented the efficacy of adequate analgesia in combination with IS in preventing ACS in patients admitted for vaso-occlusive crisis. Pain control prevented splinting and resulted in improved aeration and prevention of atelectasis. Other treatments either are empiric or their effectiveness has been inferred from studies that involved other complications of SCD (ie, transfusions and exchange transfusions after stroke).[1–7] At our own institution, we found that more than half of all children who received red-cell exchange transfusion for ACS did not show evidence of ACS at presentation, and the median time to exchange transfusion was 1 day.[23] In addition, we could detect no consistent criteria for performing an exchange transfusion.

In this article we discuss the development and, more importantly, the implementation of an evidence-based guideline for children and adolescents with SCD with ACS or at risk for ACS. The guideline was based on interventions to potentially decrease the development of ACS in those at risk, heightened vigilance for ACS detection, and aggressive treatment to prevent significant morbidity and mortality. We embarked on an implementation program, because data in the pilot study indicated that average length of stay had not been affected and pulmonary interventions and clinical respiratory score (CRS) assessments were not being administered according to the guideline. Outcome measures designed to evaluate success of the guideline included length of stay, number of exchange transfusions, average cost per SCD admission, and adherence to respiratory interventions. These outcomes indirectly measured morbidity and the efficiency of our practice. Implementation of this guideline demonstrated that even when evidence is limited, standardization of care can produce significant improvement in clinical outcomes.

METHODS

Guideline Development

Physicians from hematology, emergency medicine, critical care, and pulmonary medicine along with nursing and respiratory care served as content experts to establish the clinical practice guideline. Guideline development followed the standards established by the hospital's evidence-based outcomes center, which included review preparation, exploring existing internal and external guidelines, searching for relevant evidence from the past 15 years, critically analyzing research studies, and summarizing the evidence by preparing the guideline, order sets, and an interdisciplinary plan of care. Three systematic reviews from the Cochrane Database, 5 randomized clinical trials, 29 nonrandomized studies, and 16 review articles were used in the development of the guideline. Content experts met monthly over the course of 1 year to explore the evidence and establish critical components of the guideline.

Critical Components of the Guideline

Clinical history, physical examination, oxygen saturation level, and chest radiography were used to diagnose ACS.[1–5,7–9] Here we used a broad clinical definition of ACS (ie, lower respiratory symptoms including hypoxemia or new lung infiltrate) rather than the narrower definition used for epidemiologic or clinical studies. Essential components of the history and examination are listed in Table 1. To achieve standardization of respiratory assess-

TABLE 1 History and Physical Examination: ACS

History: assess for
Lower respiratory symptoms (cough, wheezing, tachypnea, chest pain)
Fever
Previous history of ACS episode
Physical examination:
Assess respiratory rate, use of accessory muscles, color
Auscultate lungs
Obtain pulse oximetry; detect new-onset hypoxemia
Rate severity of ACS according to CRS (see Table 2)

TABLE 2 Clinical Respiratory Score

Assess	CRS		
	0	1	2
Respiratory rate (>12 mo)	<30	30–40	>40
Auscultation	Good air movement, scattered wheezing (only expiratory), loose crackles	Depressed air movement, inspiratory and expiratory wheezes	Diminished or absent breath sounds, severe wheezing, or marked prolonged expiration
Use of accessory muscles	Mild to no use of accessory muscles; mild to no retractions or nasal flaring on inspiration	Moderate intercostals retractions, mild-to-moderate use of accessory muscles, nasal flaring	Severe intercostals and substernal retractions, nasal flaring
Mental status	Normal to mildly irritable	Irritable, agitated, restless	Lethargic
Room-air oxygen saturation, %	>95	90–95	<90
Color	Normal	Pale to normal	Cyanotic, dusky

ment, we integrated a CRS to measure the severity of respiratory distress. The CRS ranged from 0 to 12 and incorporated evaluation of the patient's respiratory rate, ausculation of the lung fields, use of accessory muscles, mental status, room-air oxygen saturation, and skin/mucosal color (Table 2). Reliability of the CRS was first established with 314 children, aged 1 to 18 years, who presented to the emergency center with symptoms that suggested asthma.[24] Before implementing the new guideline, the CRS was examined in 73 children who presented to the emergency center with a diagnosis of SCD. The mean age of this group was 12 years. The Respiratory Distress Assessment Instrument (RDAI) was used to obtain a score of respiratory severity for comparison with the CRS each time it was assessed. The RDAI is a well-published scale that is similar in assessment categories to the CRS and measures adventitious lung sounds on expiration and inspiration and retractions observed in the supraclavicular, intercostal, and subcostal spaces. In the 73 children with SCD, there was a significant ($P = .004$) association between the CRS and RDAI and the final disposition of the patients (being hospitalized or sent home). The receiver operating characteristic (ROC) curve was computed to evaluate accuracy of the CRS. The CRS at time of discharge from the emergency center was more sensitive in predicting discharge (ROC curve: 0.63) compared with the RDAI (ROC curve: 0.51). Test-retest reliability of the CRS instrument was determined by having a second health care provider rate a subset of patients with SCD independently within 15 minutes of each CRS measurement and then computing the correlation analysis. CRS correlations among 2 respiratory therapists who rated children at the time of emergency-center admission ($P < .001$), before treatment ($P < .001$), and after treatment ($P < .001$) were significant, which supports the scale as an effective clinical tool that can be used consistently by health care providers.

Respiratory therapists and physicians used the CRS to evaluate children with SCD who presented to the emergency center with respiratory illnesses and those at risk for developing ACS (fever, pain, vaso-occlusive disease). The CRS was used to guide treatment for all patients with SCD (Table 2). All patients admitted on the ACS/SCD guideline received IS alternating with PEP (an airway-clearance device) therapy every 2 hours from 8 AM until 10 PM each day. Therapy continued beyond 10 PM if the patient was awake (Table 3). Patients also received supplemental oxygen, if needed, to maintain oxygen saturations of ≥94% (Table 3). The IS and PEP were coordinated between nursing and respiratory care. Every 4 hours the patient was assessed by using the CRS, and the score was documented on the flow sheet (Table 3). Respiratory interventions and the trigger for their implementation are outlined in the clinical algorithm (Fig 1) and are based on patient CRS.

In addition to CRS assessment and administration of respiratory interventions, the clinician's role included prompt interventions based on the clinical algorithm. Ensuring that pain was adequately managed, monitoring intake and output to prevent fluid overload, administering antibiotics as indicated, monitoring laboratory results, alerting the physician team if the hemoglobin transfusion threshold had been reached, and ambulating patients at least twice daily were important care strategies (Table 3). Nurses and respiratory therapists were responsible for communicating with the medical team when CRSs escalated and assisting with patient transfers to a higher level of care as needed. Children with wheezing or rales/crackles present received specific attention, including steroid administration (Fig 1). Discharge criteria also were established to avoid unnecessary variations in length of stay (Table 4). All patients with recurrent or severe ACS were referred to a SCD pulmonary clinic.

In addition to the guideline, physician order sets were developed for use in the emergency center and outpatient hematology clinic and on the

TABLE 3 Essential Management Components for Children With SCD at Risk for ACS and Those With ACS

	Recommend	Consider
Monitoring	Vital signs (blood pressure, heart rate, respiratory rate) every 4 h Pulse oximetry every 12 h or continuous (encouraging ambulation) Weigh daily Strict charting of fluid intake/output Pain-severity rating, minimum every 4 h Monitor CRS at least every 4 h (see Table 1)	—
Diagnostics	Complete blood count, differential, platelet count, and reticulocyte count (initially and daily until improved; compare with patient's baseline values)	If history of recent fever, consider blood culture
	Chest radiograph if cough, chest pain, hypoxemia, or any respiratory symptoms are present or develop after admission	If severe illness or hemoglobin level >1 g/dL below baseline hemoglobin level, strongly consider type and cross-match If severe illness, consider obtaining serum urea nitrogen, creatinine, alanine aminotransferase, aspartate aminotransferase, and bilirubin levels If severe abdominal pain, consider abdominal ultrasound
Fluids, nutrition, general care	Total fluid intake (intravenous plus oral) at 1× maintenance	If patient is dehydrated or insensible losses are increased as with persistent fever, administer more fluids
	Encourage ambulation, activity	If patient is <5 y old: consider introducing the use of IS and PEP depending on cooperation
	If patient is ≥5 y old, use IS as follows: supervised IS, 10 breaths every 2 h between 8 AM and 10 PM, and alternate the use of PEP; IS should be documented alternately by respiratory therapist and nursing services	Consider making the patient blow soap bubbles If signs of fluid overload: intravenous furosemide 0.5 mg/kg ×1
Oxygen	Maintain pulse oxygenation at ≥94% Notify care providers of increased fraction of inspired oxygen requirement or increase in CRS	—
Pain management	Acetaminophen: 15 mg/kg orally every 4 h or as needed for temperature at >38°C; maximal daily dose: 75 mg/kg per d Morphine: 0.05–0.15 mg/kg intravenously every 2 h or 0.05–0.10 mg/kg per h continuous infusion or patient-controlled analgesia Ibuprofen (if no contraindications): 10 mg/kg orally every 6–8 h; limit frequent dosing to 72-h duration	—
Antibiotics	Discontinue prophylactic penicillin while on wide-spectrum antibiotics Add azithromycin: 10 mg/kg orally on day 1 (maximum: 500 mg per dose), then 5 mg/kg orally once daily (maximum: 250 mg per dose) for 4 d (erythromycin may be substituted)	If febrile, cefotaxime: 50 mg/kg per dose every 8 h (maximum: 1–2 g per dose) If patient is allergic, substitute clindamycin If severe illness, Consider addition of (maximum dose: 1 g) every 8 h intravenous vancomycin at 10–15 mg/kg (maximum dose: 1 g) every 8 h

acute care floors. The essential components of the diagnoses and management of children at risk for ACS and those with ACS are outlined in Table 3. Note that those children at risk for ACS were monitored for development of ACS and received interventions to potentially decrease its development.

Guideline Implementation

The specific aim of implementation was to improve utilization and compliance with the ACS/SCD guideline. An implementation team, comprising members of the initial content expert guideline development team, was formed. The team first met to assess barriers to implementation and establish outcome measures. Assessing potential barriers allowed the team to develop more targeted, effective interventions. Barriers identified included (1) lack of physician awareness of the clinical guideline, (2) nonengagement among the specialties and disciplines, (3) lack of availability of the paper physician order set, and (4) uncertainty regarding what service was responsible for administering respiratory interventions. Measures identified for gauging the success of implementation included (1) average length of stay, (2) number of exchange transfusions, (3) average cost per SCD admission, and (4) documentation of the CRS and pulmonary interventions. The implementation team used the Institute

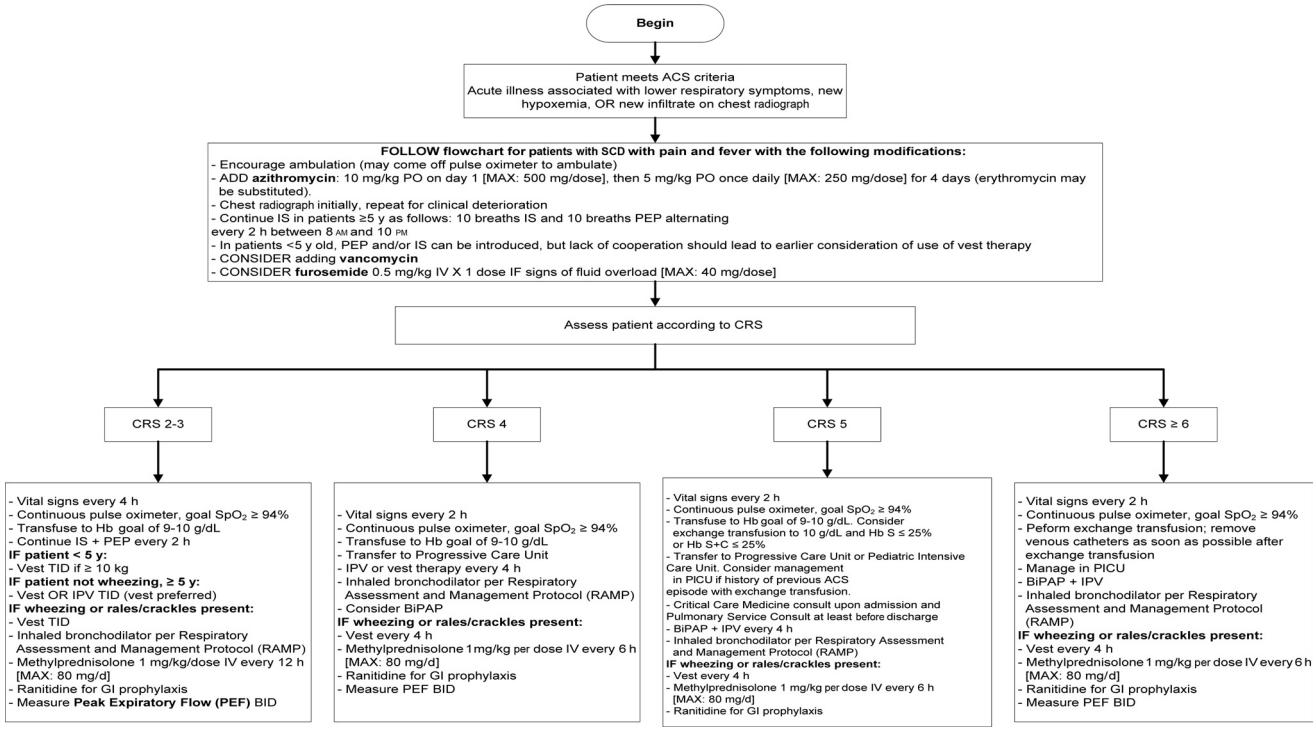


FIGURE 1

Clinical algorithm for ACS respiratory support management. PO indicates by mouth; IV, intravenous; $SP0_2$, oxygen saturation; IPV, intrapulmonary percussive ventilation; BID, twice daily; TID, three times daily; GI, gastrointestinal; Hb, hemoglobin; BiPAP, bilevel positive airway pressure.

for Healthcare Improvement's plan-do-study-act (PDSA) model to document a test of change with the ACS/SCD guideline.[25,26] PDSA is a systematic approach to problem-solving and a 4-step process for continuous quality improvement. Interventions and data from 2 PDSA cycles (phases 1 and 2) are summarized below. Pilot data were collected before the onset of the first PDSA cycle. Pilot data provided the implementation team with baseline measures to compare with future assessments after the guideline was implemented. As a result of the pilot data, specific barriers to implementation were identified, and an education initiative was launched to address these issues.

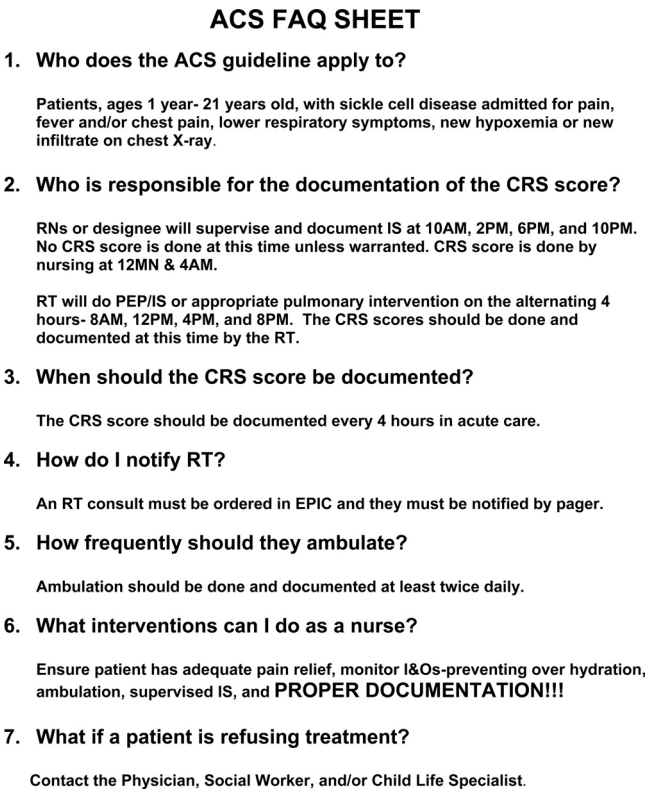
TABLE 4 ACS/SCD Discharge Criteria

Improved pulmonary symptoms and documentation of adequate oxygenation on room air
Afebrile for ≥24 h and negative cultures for ≥24–48 h, if applicable
Stable hemoglobin/hematocrit levels
Taking adequate oral fluids and able to take oral medications, if applicable
Adequate pain relief, if needed, with oral analgesics
Slow wean from steroids (over 7–10 d)
Parent understands
Discharge care
When hematology/oncology and pulmonary SCD clinic follow-ups are scheduled
Home care agencies notified as needed

Education

Phase 1/PDSA 1

The ACS/SCD guideline was designed for use by multiple specialties throughout the hospital. All specialties routinely caring for patients with SCD were identified from the pilot data, and education sessions were developed for each specialty. Individualized in-services were held for staff from the emergency center, inpatient hematology-oncology unit, general medicine, float pool staff, progressive care unit, outpatient hematology clinic, and respiratory care. Physicians, fellows, and residents who work in these specialty areas were educated on the ACS/SCD guideline through noon conferences, education modules, lectures, educational lunches, and grand rounds. Presentations provided a brief description of ACS and its complications, reasons for implementing the guideline, and bullet points with information pertinent to the physician, nurse, and respiratory therapist roles in caring for patients with SCD. Case scenarios were presented to allow staff to apply the guideline to real-life situations. A frequently-asked-questions hand-out was given to use as a quick reference guide (Fig 2). In addition, a video describing pulmonary pathophysiology, and the rationale for the guideline's pulmonary interventions, was developed and placed on the hospital's intranet site to augment live in-services.



ACS FAQ SHEET

1. **Who does the ACS guideline apply to?**

   Patients, ages 1 year- 21 years old, with sickle cell disease admitted for pain, fever and/or chest pain, lower respiratory symptoms, new hypoxemia or new infiltrate on chest X-ray.

2. **Who is responsible for the documentation of the CRS score?**

   RNs or designee will supervise and document IS at 10AM, 2PM, 6PM, and 10PM. No CRS score is done at this time unless warranted. CRS score is done by nursing at 12MN & 4AM.

   RT will do PEP/IS or appropriate pulmonary intervention on the alternating 4 hours- 8AM, 12PM, 4PM, and 8PM. The CRS scores should be done and documented at this time by the RT.

3. **When should the CRS score be documented?**

   The CRS score should be documented every 4 hours in acute care.

4. **How do I notify RT?**

   An RT consult must be ordered in EPIC and they must be notified by pager.

5. **How frequently should they ambulate?**

   Ambulation should be done and documented at least twice daily.

6. **What interventions can I do as a nurse?**

   Ensure patient has adequate pain relief, monitor I&Os-preventing over hydration, ambulation, supervised IS, and **PROPER DOCUMENTATION!!!**

7. **What if a patient is refusing treatment?**

   Contact the Physician, Social Worker, and/or Child Life Specialist.

FIGURE 2
Frequently-asked-questions (FAQ) hand-out provided to inpatient nurses caring for patients with SCD with ACS or at risk for ACS.

Phase 2/ PDSA 2

Initial evaluation of guideline usage after implementation of the education initiative revealed a deficit in documentation of both the CRS and pulmonary interventions. The second PDSA cycle focused on improving documentation of these 2 components. Small group meetings with nursing and respiratory care representatives from the implementation team were held. Data from the first PDSA cycle were shared with nursing and respiratory care educators and managers, who then disseminated the data to their colleagues, highlighting opportunities for quality improvement. The guideline-implementation coordinator also presented the data and solicited feedback from nurses at shift report on all inpatient units. In addition, CRS badge cards with the CRS scale and bulleted points essential to the ACS/SCD guideline were given to nurses on the 3 units that received the highest volume of children admitted with SCD.

Methods of Evaluation

Pilot Study

A retrospective chart review was performed for all guideline-eligible patients (all patients from 1 to 21 years of age admitted with SCD) from February through March of 2009. Data on length of stay, number of exchange transfusions, and documentation of pulmonary interventions and CRS assessments for the first 3 days of admission were collected for each SCD admission.

Phase 1/PDSA 1

A data-collection form was developed to capture outcome measures established by the implementation team for the 2 subsequent PDSA cycles. A retrospective chart review was performed for all guideline-eligible patients from October through November of 2009. Data on length of stay and number of exchange transfusions over the entire inpatient stay for each patient with SCD were also collected. Data on the average cost per patient admission for SCD were obtained from a financial analyst who searched a database by using All-Patient Refined Diagnosis-Related Groups codes.

Data on documentation of CRS and pulmonary interventions for the first 3 inpatient days for every patient with SCD were collected. As indicated in the guideline, the CRS was to be assessed every 4 hours and supervised IS/PEP therapy administered every 2 hours between the hours of 8 AM and 10 PM. These responsibilities were shared between nursing and respiratory care. The number of opportunities for nurses/respiratory therapists to document the CRS and pulmonary interventions administered was compared with the number of times there was actual documentation of the CRS and pulmonary interventions performed.

Phase 2/PDSA 2

Identical methods of evaluation were used for the second PDSA cycle, because the same outcome measures were monitored. Data on patient admissions for SCD from June through August of 2010 were collected for the second PDSA cycle.

In addition to the data collected in the 2 PDSA cycles, data on average length of stay for patients with SCD were evaluated in fiscal years 2007, 2008, and 2009 and the first 6 months of fiscal year 2010. These data were obtained from a financial analyst who searched a database by using All-Patient Refined Diagnosis-Related Groups codes.

We did not obtain institutional review board approval for this retrospective analysis, because it was a quality-improvement exercise, not a research study.

Analysis

Minitab statistical software (Minitab Inc, State College, PA) was used to analyze the data. A 2-sample *t* test was run to evaluate the difference in length of stay between the pilot data and each of the 2 PDSA cycles. A 2-proportions test was run to assess if there was a difference in documentation of the CRS and pulmonary interventions between the initial pilot data and the data collected after the interventions in each of the 2 PDSA cycles.

RESULTS

The guideline was developed and instituted in February 2008 after informal education interventions with nursing, respiratory care, and physician groups. The average length of stay did not appreciably change from pre–guideline development in 2007 to up to 22 months after institution of the guideline. The formal implementation team was initiated in the summer of 2009.

Three measurement periods were used to evaluate the formal implementation of the ACS/SCD guideline. In the pilot study conducted from January to February of 2009, 1 year after the guideline was completed and instituted, 59 patients older than 1 year with SCD and at risk for ACS or with ACS were admitted. Fifty-five charts were available for review; 1 patient with a prolonged length of stay of 38 days was removed from analysis, because the extended hospitalization was unrelated to ACS. Sixty-two patients older than 1 year were admitted from October to November of 2009; 61 charts were available for review for the first PDSA cycle evaluation, because 1 patient had a prolonged length of stay of 25 days and was not included because the extended hospitalization was unrelated to ACS. Twenty-four patients older than 1 year were admitted from June through August of 2010, and all were reviewed and included in the analysis of the second PDSA cycle.

The discharge criteria facilitated decision-making at the time of discharge and decreased length of stay by standardizing the approach to discharge. Average length of stay was reduced from 5.8 days during the pilot study before implementation to 4.1 days after the second PDSA cycle, which is a reduction of 1.7 days (29%) (95% confidence interval for difference: 0.1–3.2) (Table 5). No patients in the pilot study or the 2 PDSA-cycle evaluations received an exchange transfusion. Average cost per SCD admission decreased from $30 359 in fiscal year 2008 before implementation of the guideline to $22 368 in the first 6 months of 2010, which is a reduction of $7991 (26.3%). On average, this hospital admits 357 patients per year for SCD. The reduced average cost per admission would result in an annual savings of more than $2 850 000.

Documentation of the CRS increased from 31% before implementation of the guideline to 75.5% after the second PDSA cycle, which is an improvement of 44.5% (95% confidence interval for difference: 38.7%–50.2%). Documentation of IS/PEP increased from 23% before implementation of the guideline to >50% after the second PDSA cycle, which is an improvement of 27% (95% confidence interval for difference: 21.3%–33.0%) (Table 5). Patients with a higher CRS in the emergency center were more likely to receive a simple blood transfusion while hospitalized ($P < .001$). CRS in the emergency center was not related to the patient's length of stay or transfer to the critical care unit.

Average length of stay for patients with SCD decreased from 5.4 days in 2007, 2008, and 2009 to 4.1 days in the first 6 months of fiscal year 2010.

DISCUSSION

The mainstay of successful treatment for children with SCD with ACS or at risk for ACS is high-quality supportive care. Implementation of a standardized approach to care for children at risk for ACS and those with ACS revealed significant changes in clinical outcomes. Standardizing fluid management, maintaining oxygenation, administering respiratory interventions, and ensuring pain control were essential elements of quality care for these patients. We also instituted an aggressive respiratory monitoring and therapy program. In addition, using the CRS afforded us a common language for describing the condition of these children to each other and driving treatment and triage. The use of the CRS as a standardized method for evaluating clinical respiratory status played an important role in minimizing variation in assessment as well as management for these children.

The decision whether to institute an exchange transfusion, and determinants of the need for acute respiratory moni-

TABLE 5 Outcome Measures at Baseline, Phases I and II

Outcome Measures	Baseline, Jan–Feb 2009 ($N = 54$)	Phase I, Oct–Nov 2009 ($N = 61$)	Phase II, June–Aug 2010 ($N = 24$)
Average length of stay, d	5.8	4.3	4.1
Exchange transfusions, *n*	0	0	0
Hospitalization cost, US $	30 359	—	22 368
Opportunities to document CRS, *n*	831	866	310
CRS documented, *n* (%)	258 (31.0)	491 (56.7)	234 (75.5)
Improvement in CRS documentation, % (*P*)	—	25.7 (<.001)	18.8 (< .001)
Opportunities to document IS, *n*	974	1197	353
IS documented, *n* (%)	227 (23.3)	418 (34.9)	178 (50.4)
Improvement in IS documentation, % (*P*)	—	11.6 (<.001)	15.5 (<.001)

toring, were inconsistent and undefined before the guideline's development. On the basis of results from our earlier study,[23] it is remarkable that no exchange transfusions were needed for children with ACS during the 3 monitoring periods. Thus, we believe a standardized approach to exchange transfusion resulted in decreased use of this therapy during the initial roll-out of the protocol even before the guideline was formally implemented. One explanation is the fairly rapid adoption of the guideline by our critical care service.

It is important to recognize the critical role of implementation in improving patient outcomes. The guideline was formally implemented in the summer of 2009, more than 1 year after the guideline's development and dissemination. It was only after implementation that patient outcomes improved. Data showed the step-wise improvement in outcomes from the pilot data collected before implementation through each of the 2 implementation phases (Table 5).

The change in average cost per SCD admission is impressive. This decrease is most likely attributable to the decrease in average length of stay and room charges for SCD admissions after implementation of the guideline.

One of the resources required for implementation included a master's level trained implementation coordinator. This person coordinated implementation programs for several evidence-based guidelines and devoted approximately one-third of her time to this project. In addition, the implementation team was vital to the project's success. The team comprised ~15 members representing various medical specialties, nursing, respiratory care, and other allied health professions. Team members attended several meetings to assess barriers and define outcome measures, participated in the education initiative, and served as champions for the project by engaging their colleagues in discussions about the guideline.

Although our institution serves one of the largest populations of people with SCD in the country and has a highly comprehensive SCD center, we feel that one of the exciting things about this project is its ability to be replicated. The success of the program rested heavily on education and creating awareness. None of the interventions implemented were terribly sophisticated or required a great deal of resources. The guideline and CRS provided the framework for standardizing care. A smaller, more resource-constrained institution should also see improvements in clinical outcomes for patients if they have engaged providers who are willing to implement such a framework. It may be easier, in fact, for smaller institutions to implement such a program, because their size would enable them to develop more targeted interventions.

Although we saw improvements in outcomes for patients with SCD, we identified a number of barriers to and opportunities for quality improvement after the second and final PDSA cycle. Documentation of the CRS and IS was poorer on those units that received a lower volume of admissions for SCD. In addition, nurses documented less than respiratory care providers on all units, which may have been, in part, because of the chart design. Nurses now are required to document in 2 different places. As our institution goes live with computerized physician order entry and electronic chart documentation this year, one of our next PDSA cycles may include examining ways to improve documentation by modifying the nursing flow chart in our institution's electronic medical record. These issues highlight the fact that quality improvement is an ongoing process.

There are limitations to this quality-improvement project. The number of charts reviewed in the second phase of implementation was significantly less than the number reviewed in the pilot study and the first phase of implementation. There were fewer SCD admissions during this period. We could not detect the specific mechanisms by which implementation improved outcomes; thus, we do not know which portions of the guideline may be critical to its success.

It is unlikely that the outcomes of this quality-improvement project were influenced by other systematic changes during the implementation period. The institution's practices concerning discharge did not change after the guideline was developed. In addition, there was no difference in severity of ACS presentations over the course of the guideline's development and implementation. We do not have data on the use of hydroxyurea during this period. It is possible that an increase in the use of this drug in the population of patients with SCD had an impact on clinical outcomes, but we feel that the reduction in average length of stay can be attributed to both improvements made in the administration and documentation of the CRS and pulmonary interventions and the avoidance of exchange transfusions.

CONCLUSIONS

Standardization of care improves clinical outcomes for patients with SCD who are at risk for ACS or have ACS. It is even more important to note that standardization of care did not result from guideline development alone but also from implementation efforts. We advocate for, and encourage other providers of patients with SCD to adopt, an evidence-based model of care and to devote the resources necessary for implementation. The results of this project indicate that both components are vital for improving the clinical outcomes of patients with SCD.

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Pediatrics 2011;127;e480; originally published online January 17, 2011;
DOI: 10.1542/peds.2010-3099