Positive Expiratory Pressure Device Acceptance by Hospitalized Children With Sickle Cell Disease Is Comparable to Incentive Spirometry

Lewis L Hsu MD PhD, Brenda K Batts MPH RRT, and Joseph L Rau PhD RRT FAARC

BACKGROUND: The pulmonary complication in sickle cell disease known as acute chest syndrome (ACS) has potential for high morbidity and mortality. A randomized trial demonstrated that incentive spirometry (IS) reduces the rate of ACS, leading to a role for respiratory therapy in hospital management of sickle cell pain. However, use of IS can be limited by chest wall pain, or by difficulty with the coordinated inspiration in a young child. Intermittent positive expiratory pressure (PEP) therapy may be easier for a child's coordination and more comfortable than IS for a child with chest wall pain. PURPOSE: To compare PEP therapy with conventional IS for children hospitalized for sickle cell pain with respect to patient satisfaction, length of hospital stay, and progression to ACS. METHODS: This pilot study enrolled 20 children upon hospitalization for sickle cell pain in the thorax, randomly assigning them to either PEP ($n = 11$) or IS ($n = 9$) therapy, administered by a therapist hourly while awake. RESULTS: The randomization assigned an older distribution to PEP than IS (12.3 vs 8.8 y). Patient satisfaction was high for both respiratory care devices, and there was no difference between the PEP and IS groups (4.5 vs 4.4, $p = 0.81$). Length of hospital stay was similar (5 vs 4.3 d, $p = 0.56$). No children in either group progressed to ACS. CONCLUSION: These preliminary results show no difference in the primary outcomes in the 2 groups. Intermittent PEP therapy warrants further study as an alternative to IS for sickle cell patients at high risk for ACS, as effective preventive respiratory therapy. *Key words: sickle cell, intermittent positive expiratory pressure, incentive spirometry, child, patient satisfaction, acute chest syndrome, respiratory therapy, hospital pain management.* [Respir Care 2005;50(5):624–627. © 2005 Daedalus Enterprises]

Introduction

The pulmonary complication in sickle cell disease known as acute chest syndrome (ACS) has potential for high morbidity and mortality.[1–3] ACS is defined as a new radiographic opacity (nonatelectatic) in a patient with sickle cell disease, who has an additional chest symptom such as cough, fever, or chest pain.[1–2] Multiple etiologies can contribute to ACS (pneumonia, atelectasis, rib infarcts, marrow fat embolism, and lung infarct) and lead to pneumonitis with vaso-occlusion that can be rapidly fatal.[1] Children with sickle cell pain in the thorax are at high risk for ACS due to splinting respirations, but a randomized controlled trial of incentive spirometry (IS)[4] reduced the rate of ACS in such children from 8/19 to a rate of 1/19. Its presumed mechanism for preventing ACS is reducing mismatch of

Lewis L Hsu MD PhD is affiliated with Hughes Spalding Children's Hospital, Atlanta, Georgia, and with the Division of Pediatric Hematology, Drexel University College of Medicine, Philadelphia, Pennsylvania. Brenda K Batts MPH RRT is affiliated with Hughes Spalding Children's Hospital, Atlanta, Georgia. Joseph L Rau PhD RRT FAARC was affiliated with the Department of Cardiopulmonary Care Sciences, Georgia State University, Atlanta, Georgia, at the time of this study; he is now Professor Emeritus.

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Correspondence: Lewis L Hsu MD PhD, Pediatric Hematology, St Christopher's Hospital for Children, Drexel University College of Medicine, Erie Avenue at Front Street, Philadelphia PA 19134. E-mail: lhsu@mail.nih.gov.

regional ventilation and perfusion, and reducing atelectasis. IS has gained acceptance as an adjunct to hospital management for sickle cell pain, as demonstrated by inclusion in clinical pathways and guidelines.[5–7] However, use of IS can be limited by chest wall pain or by difficulty with the coordinated inspiration in a young child. Positive expiratory pressure (PEP) devices reduce atelectasis by offering an expiratory retard during a normal breathing cycle, which maintains increased lung volumes during a lengthened expiratory phase and prevents airway collapse. Intermittent PEP therapy may be easier for a child's coordination and more comfortable than IS for a child with chest wall pain. Intermittent PEP is also less costly than continuous positive airway pressure. Only one randomized comparison of PEP and IS has been published. Sanchez et al[8] examined 27 cystic fibrosis subjects in a cross-over trial and found IS was superior in improving peak expiratory flow, but symptom scores were no different between IS and PEP.

The purpose of the present study was to compare PEP therapy with conventional IS for children hospitalized for sickle cell pain, in patient satisfaction and in preventing pulmonary complications and progression to ACS.

Methods

Patients

Twenty children with sickle cell disease were enrolled from 2000 to 2002, upon hospitalization for sickle cell pain in the thorax (chest wall or back) at Hughes Spalding Children's Hospital, Atlanta, Georgia. Children with an infiltrate on chest radiograph on admission were excluded, because this study focused on progression to ACS. Other exclusion criteria were age < 6 years or cognitive delay. With informed consent, subjects were randomly assigned to either PEP ($n = 11$) or IS ($n = 9$) therapy on admission. All were on room air, except for 2 in the PEP group on a small amount of nasal cannula supplemental oxygen (fraction of inspired oxygen $= 0.26$).

Equipment

Incentive spirometry was administered using a Coach 2 for Kids device (DHD Healthcare, Wampsville, New York), while PEP therapy utilized the TheraPEP device (DHD Healthcare, Wampsville, New York). All treatments were administered by a therapist every hour while awake, and continued until the patient's discharge from the hospital or progression to ACS.

Each PEP breathing maneuver consisted of the patient inhaling normally and then exhaling through a mouthpiece and a restrictor size of 1 to 4, which prolongs exhalation over a period of approximately 2 or 3 times normal, while maintaining the TheraPEP pressure indicator at 10–20 cm H_2O. This was repeated for a total of 10 breaths. The IS maneuver consisted of encouraging patients to take maximum inspiratory volumes for 10 breaths every hour while awake.

Table 1. Baseline Characteristics of Patients on Admission for Sickle Cell Pain*

Group	PEP ($n = 11$)	IS ($n = 9$)
Age	12.3 ± 1.1	8.8 ± 3.2
Sex	male = 4	male = 4
RR (breaths/min)	23 ± 3.9	24 ± 5.8
T (°C)	36.8 ± 0.4	36.0 ± 3.5
S_{pO_2} (%)	98.6 ± 1.5	97.2 ± 1.9

*Data are presented as mean ± SD. The 11 on positive expiratory therapy and the 9 assigned to incentive spirometry were similar in respiratory rate (RR), temperature (T), and blood oxygen saturation measured via pulse oximetry (S_{pO_2}). The randomization resulted in an older age distribution in the positive expiratory pressure (PEP) group than in the incentive spirometry (IS) group (mean 12.3 vs 8.8 y, SD 3.7).

Outcome Measures

Primary outcome measures were (a) patient satisfaction with therapy and (b) length of hospital stay. Children rated their satisfaction with the respiratory therapy device on a Likert scale (0 = completely unsatisfied, 5 = completely satisfied). Progression to ACS was an end point defined by new infiltrate on chest radiograph plus respiratory symptoms.[1–2]

Statistical Analysis

This pilot study was designed to detect only large differences between PEP and IS, because only large effect sizes would be clinically relevant in this very heterogeneous disease. The sample size of 20 had a power of 88% to detect a difference between treatment groups of 2 days of hospitalization, or 15% difference on the Likert scale, at a significance level of 0.05 for comparisons by unpaired 2-tailed *t* test. The largest differences that could have been missed were estimated as 1.87 days and 12% difference on the Likert scale. A 1-sided *t* test would compare whether PEP was superior to IS, but would have provided similar sensitivity, missing the largest differences of 1.6 days and 8% difference on the Likert scale.

Results

The PEP and IS groups had similar physiologic measures upon hospital admission (Table 1), although 2 in the PEP group had chronic mild hypoxia and received 26%

oxygen on admission. The randomization assigned an older distribution to PEP than IS (12.3 vs 8.8 y).

Patient satisfaction on the 5-point Likert scale was high and not significantly different for the 2 groups. Mean and SD were comparable for the use of PEP and IS: 4.5 ± 0.5 and 4.4 ± 0.5, p = 0.81 by 2-tailed *t* test. The 95% confidence intervals of means were 4.0 to 4.9 and 4.0 to 4.8, respectively.

Length of hospital stay also was similar for the 2 groups: mean and SEM were 5.0 days ± 2.5 for positive expiratory therapy device (PEP) and 4.3 days ± 2.6 for IS, p = 0.56 by 2-tailed *t* test. The 95% confidence intervals of means were 3.3 to 6.7 and 2.4 to 6.3, respectively.

No children in either group progressed to ACS. None dropped out of the study.

Discussion

This pilot study found no significant differences between PEP and IS in satisfaction scores and length of hospitalization. This study suggests that PEP may be equivalent to IS for children hospitalized with sickle cell pain, but several limitations of the study should be mentioned.

Limitations

This study's randomization may actually have underestimated the effectiveness or satisfaction with PEP, because the PEP group was older and included 2 chronically hypoxic patients. Younger children may find PEP easier to use than IS. A larger study in the future may need to stratify by age and/or baseline oximetry, or focus on younger children.

The principal limitation was the small scale of this pilot study, which was designed with sufficient statistical power to detect only large differences between the 2 treatment groups (2 d length of hospital stay or 15% difference on the Likert scale). Smaller differences between treatment groups may have been missed. However, smaller differences may not be clinically relevant.

None of the 20 children developed ACS in this study, despite being at high risk due to sickle cell pain in the chest wall. This rate of ACS was similar to that described by Bellet et al,[4] in which IS reduced the rate of ACS to 1 out of 19 patients at risk, compared to 9 of 19 children not using IS. Determining whether PEP is superior to IS in preventing ACS, or detecting a 50% difference in the rate of ACS from the Bellet et al study, would have required over 470 subjects (chi-square comparison of proportions, degrees of freedom = 1).

An additional limitation of the present study is a lack of objective pulmonary function measures, including lung volumes. Obtaining such measures on a younger population can be difficult, especially in the presence of sickle cell crisis and pain. Both PEP and IS maneuvers were intended to recruit alveolar ventilation, but the effect on lung mechanics differ and the 2 maneuvers might provide a different pattern of benefit on pulmonary function. Confirmation that PEP and IS maintained maximal lung volume would have been helpful, but this pilot study was not able to include spirometry or plethysmography.

Finally, the positive effect of PEP and IS may be simply from frequent personal attention by a respiratory therapist, regardless of whether the patient achieves maximal lung volume maneuvers. In an early study of respiratory care to prevent pulmonary complications in adults after upper abdominal surgery, Stock et al speculate:

> . . . the low incidence of pneumonia regardless of the type of therapy may be attributable to vigorous, vigilant respiratory care in a population at high risk for developing pneumonia. . . . Frequency and supervision of respiratory therapy may be more important than the type of therapy delivered.[9]

It is possible that respiratory therapists play a similar key role in sickle cell care and provide a basis for the use of either PEP or IS to reduce the high risk of ACS and for the therapeutic impact of respiratory therapy in this blood disorder.

Conclusion

This study is the first to explore respiratory therapy in sickle cell disease since the demonstration of the dramatic benefit of IS in reducing the rate of ACS.[4] The results of this small study suggest that PEP therapy was comparable to conventional IS for children hospitalized with sickle cell pain, based on measures of patient satisfaction and length of hospital stay. Intermittent PEP therapy warrants further study as an alternative to IS for sickle cell patients at high risk for ACS.

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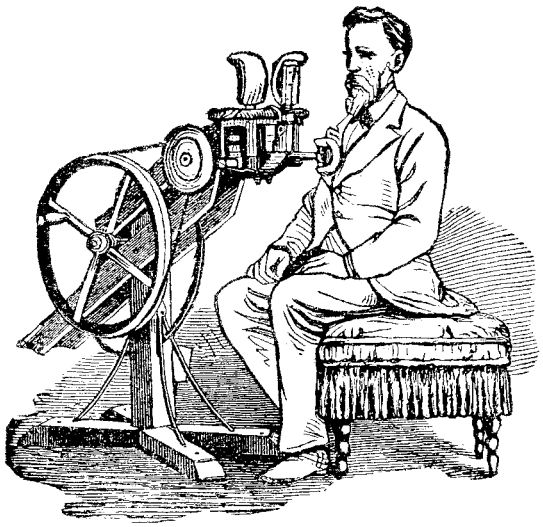

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