Home management of sickle cell-related pain in children and adolescents: natural history and impact on school attendance

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Summary

Some children and adolescents with sickle cell disease experience frequent painful episodes. To gain information about the natural history of the pain and its impact on sleep and school attendance, we developed a home-based diary system. Eighteen children and adolescents completed 4756 diary days, with an average compliance of 75%. Pain was reported on 30% of days and was managed at home nine-tenths of the time. Girls reported more days with pain than did boys, and age was positively correlated with the length of the painful episodes. The pain affected school attendance and sleep. Patients were absent from school on 21% of 3186 school days, with half of the absenteeisms on days with reported pain. Of the pain-associated absenteeisms, two-thirds occurred when pain was managed at home, and one-third when patients were hospitalized. The average consecutive number of school days missed was 2.7. These findings have implications for developmentally critical activities.

Introduction

Vaso-occlusive pain is the most common problem encountered by patients with sickle cell disease (SCD) (Tetrault et al. 1974). The pain, which is intermittent, often severe, and unpredictable, can start as early as 6–9 months of age, and continues throughout adulthood (Serjeant 1985). The pain frequency and severity varies greatly among and within individuals (Vichinsky et al. 1982; Serjeant 1985), with a minority of patients experiencing frequent and severe painful episodes (Vichinsky et al. 1982). Many studies of the natural history and treatment of SCD-related pain utilize emergency department and inpatient admissions as indicators of pain severity and frequency (Platt et al. 1991). However, patients recount that painful episodes are often managed at home. The effect of home pain management on activities necessary for normal development, such as attending school, has not been described.

Children with chronic illnesses often miss school. Public policy in many states dictates that homebound instruction is provided to ill students only after a certain consecutive number of school days are missed (Hobbs et al. 1985). The impact of SCD-related pain on school attendance and the average consecutive number of days missed has not been delineated. We report data on the frequency of sickle cell-related pain and its effects on school attendance from 18 children and adolescents followed over an average of 10 months each. The data were collected using a home pain diary developed specifically for patients with SCD (Shapiro et al. 1990).

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Methods

Patients

Patients were enrolled in a study of the natural history of sickle cell-related pain, and of the efficacy of self-hypnosis as an intervention. Eligibility criteria included (a) diagnosis of a sickle hemoglobinopathy; (b) age between 7 and 17 years; (c) retrospective report of at least 4 painful episodes over the previous year; and (d) identification of pain as a significant problem by the patient and the family. Exclusion criteria were regular red blood cell transfusion therapy, co-existence of other serious chronic illnesses, mental retardation, and emotional or family problems requiring ongoing psychotherapy.

All patients were followed at the Comprehensive Sickle Cell Center of The Children's Hospital of Philadelphia. Of the 380 patients followed at the Center, 144 were eligible by age and diagnosis, and met no criteria for exclusion. Thirty-three of these patients could not be contacted, and 78 did not have frequent enough painful episodes to meet the eligibility criteria. Nine eligible patients declined to participate, citing reasons such as "could not make appointments" (n = 5); "not interested" (n = 2); and family member did not want the patient to participate (n = 2). Six patients who expressed initial interest could not subsequently be contacted.

The enrolled patients ranged from 8 to 17 years of age (average: 13 years). There were 7 girls and 11 boys; boys and girls did not differ significantly in age. The average length of time that patients stayed in the study was 10 months, with a range of 2–15 months.

The Committee for the Protection of Human Subjects at The Children's Hospital of Philadelphia approved the study. Informed consent was obtained from parents and patients before enrollment.

Home Diary

A pain diary was developed from a self-report system used for research on sleep and circadian rhythms (Shapiro et al. 1990). The diaries were compact, each 5×7 in., and covering a 1-week period of time. Patients were asked to complete 2 pages each day, 1 in the morning as soon after awakening from sleep as convenient, and 1 in the evening before going to bed. On the morning page, patients indicated the presence of any SCD-related pain by circling 'yes' or

'no'. If 'yes' was circled, they indicated the pain intensity on a 10-cm linear analogue scale. Patients also indicated by circling 'yes' or 'no' whether they slept well, and rated their perceived sleep length on a 5-cm linear analogue scale. At night, children were again asked about the presence and intensity of SCD-related pain, school attendance on weekdays, and medication use.

The diaries were attractively printed on heavy paper. The cover page was personalized with the child's name, the dates of the week, and the statement that this was a confidential record. On the last page was a statement thanking the child for completing the diary, and reminding the patient to place the diary in the mail.

Procedures

Patients and their families were told that participation in the study involved a responsibility to complete and return the diaries. Possible difficulties in adherence were openly discussed, and a joint problem solving approach used to define solutions that were compatible with the children's and families' lifestyles and preferences. It was emphasized that responsibility for the diaries should not add stress to daily life, nor should it precipitate conflicts between parents and children. The relative responsibilities of children and their parents were defined, with individual variations based on factors such as developmental maturity and interactional styles. In general, parents helped children under the ages of 10 or 11 remember to complete the diaries, whereas older children were encouraged to develop their own reminder systems. Older teenagers took full responsibility for the entire process.

To facilitate completion and return of the diaries, procedures were instituted that had been used successfully in long-term studies of mood (Dinges 1989). Each week a packet containing the diary for the following week and a pre-addressed stamped envelope in which to return the diary was mailed to the patient. Two of the authors (B.S.S. and L.B.R.) contacted the families by telephone on an average of once every 2–3 weeks. During these calls, we discussed the entries in the diaries that we had received, provided positive reinforcement and reminders when necessary, and elicited concerns and questions about pain management and study interventions. If less than 25% of the diaries were returned for 2 months in a row, the patients and families were asked whether they still wished to be in

TABLE I

DIARY ADHERENCE, PAIN DAYS AND HOSPITALIZATION

Case	Sex	Age	Completed diary days	%Days in pain	Hospitali-zations	% Pain days in hospital
1	M	13	416	5.7	1	23.8
2	M	16	294	10.5	2	51.6
3	F	13	35	65.7	0	0.0
4	F	13	404	31.6	5	21.9
5	M	11	213	19.6	2	19.0
6	F	13	152	46.0	3	21.4
7	M	14	204	7.3	0	0.0
8	M	15	259	48.6	8	34.1
9	F	17	338	37.2	1	2.3
10	F	14	18	73.6	0	0.0
11	M	12	333	8.3	0	0.0
12	M	8	318	11.3	2	33.3
13	F	10	374	9.6	2	27.7
14	M	13	432	21.7	0	0.0
15	M	9	220	34.5	2	13.1
16	F	14	275	97.4	1	5.6
17	M	11	158	3.1	0	0.0
18	M	17	309	6.4	2	80.0
Mean (SD)		12.9 (2.5)	264 (119.8)	29.9 (27.2)	1.7 (2.1)	18.5 (21.6)

the study. If further participation was declined, the patients were dropped from enrollment.

We discovered early in the study that patients who otherwise adhered to the protocol did not complete the diaries when they were hospitalized. Therefore, the diary data pertain only to home management. However, because we followed the patients closely, we knew when and why they were hospitalized. Therefore, the data include the number and timing of pain days spent in the hospital. These data were used along with the diary data to assess the relative rates of home versus hospital management, the proportion of days with pain, and the duration and number of painful episodes.

Analysis of data

Adherence to the study protocol was determined from the number of diaries completed and returned, including time spent in the hospital. Pain was evaluated both as the proportion of days that pain was reported out of the sum of the total number of diary days completed plus hospital pain days, as well as by the total number of painful episodes managed at home, in the hospital, or both (i.e., each discrete multi-day period of pain). To evaluate the effects of pain on outcome variables, each patient's home diary data was reduced according to report of the presence or absence of pain. With this system, each patient generates only 1 data value, regardless of the amount of time in the study. Thus, for each diary dependent variable (e.g., sleep quality), data were averaged within all days in which pain was not reported and separately within all days in which pain was reported. For each dependent variable, 'pain' and 'no pain' distributions were examined for homogeneity of variance. Violations resulted in use of non-parametric statistical comparisons. Mann Whitney U statistic was used to compare pain patterns of male and female patients. Wilcoxon matched-pairs signed-ranks tests were used to evaluate differences in sleep and school attendance between days with and without pain. Spearman rank-order correlations were used for assessing the relationship between age and pain. Analyses were conducted including and excluding a 14-year-old female patient who reported pain virtually every day (case 16 in Table I). Those instances in which her exclusion changed the findings are reported in the results. Outcomes were considered statistically significant if $P < 0.05$, 2-tailed.

Results

Adherence with the daily diaries

A total of 4756 completed diary days were received from the 18 patients. Patients completed an average of 264 diary days each, although there was considerable inter-subject variability (Table I). On average, 75% of the diary days were completed each month. Adherence to the diary system was inversely related to the percentage of days on which painful episodes were reported ($r = -0.658$, $P < 0.01$), and was not related to age, sex, the number or intensity of the painful episodes, or the absolute number of completed diary days. Adherence with the diaries gradually declined over the first 10 months of enrollment. Average adherence was 90% in the 1st month and 63% in the 10th month, giving an average decrease of 2.7% each month.

Home versus hospital management of pain

If any part of a painful episode involved emergency department or inpatient hospitalization, this episode was considered to have been managed in the hospital.

Pain was managed entirely at home during 89% of the painful episodes (81% of the days with pain). However, patients varied widely in hospital contact for pain management (Table I). Six patients had no hospitalizations for pain. The remainder were hospitalized for an average of 16% of painful episodes (SD = 13.73; range: 4–50%). Comparisons of these 2 subgroups revealed few significant differences. Hospitalized children reported a greater number of painful episodes (mean = 24.2, SD = 15.7) than did non-hospitalized children (mean = 10.6, SD = 10.8; $U = 13$, $\chi^2 = 4.08$, $df = 1$, $P = < 0.05$). However, groups did not differ in the proportion of days with pain (hospitalized: mean = 23.7, SD = 16.2; non-hospitalized: mean = 29.9, SD = 31.5), in the intensity of pain managed at home and reported in the diary, or in demographic or activity (e.g., school attendance, sleep) variables.

Age-related patterns

The patterns of the painful episodes varied greatly among patients. Pain was reported on an average of 30% of all days (26% with the exclusion of patient 16; see Table I), with a range of 3–97% (median = 20.7%). On average, 2 episodes of pain were reported each month (range: 0–5), with a mean duration of approximately 4 days (range: 1–17 days). Analog pain intensity ratings (0 = none, 100 = worst) during painful episodes managed at home averaged 29, with a range from 10 to 69.

Age was examined in relation to the percentage of days with pain and the frequency, length, and average intensity of painful episodes. Older patients reported longer painful episodes (rho = 0.677; $t = 3.67$; $P < 0.01$). There was no relationship between age and the frequency or intensity of pain, or the number of painful episodes.

Sleep patterns

Ratings of sleep quantity and quality were influenced by pain. Patients reported poor sleep on an average of 43% of days with pain (SD = 32%), compared to only 3% of days without pain ($z = 3.57$; $P < 0.001$; Fig. 1). Similarly, reported sleep duration was significantly less on nights in which pain occurred relative to nights without pain ($z = 3.62$; $P < 0.001$).

Effect of pain on school attendance

Potential school days (3186) were analyzed. (Days were counted as potential school days when a daily diary report was available or when patients were hospitalized for pain management and school was in session. Days for which there were no diary data and days during which patients were hospitalized for problems not related to pain were excluded.) Patients missed school on 41% (SD = 27.8%) of the days on which they reported pain, compared to an average of 9% (SD =
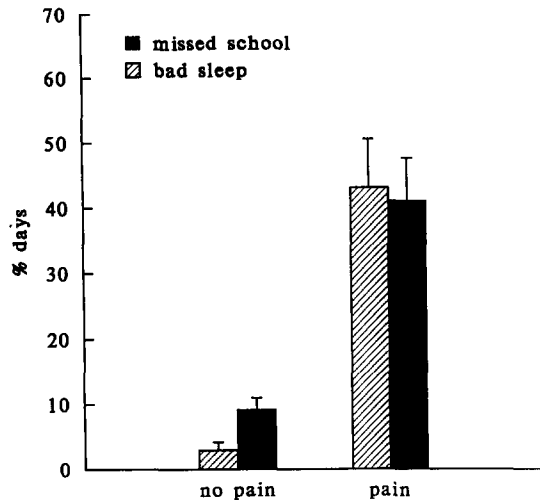
Fig. 1. Histograms of means (SEM) comparing the proportion of days without pain to the proportion of days with pain on 2 daily diary variables: reports of 'bad' sleep the night before, and reports of school absenteeism. For all variables the proportion of 'pain' days on which these outcomes occurred was significantly higher ($P < 0.001$) than the proportion of 'no pain' days on which they occurred (see text).

7.3%) on days without pain ($z = -3.29$; $P < 0.001$; Fig. 1). Of all missed school days, about one-half (11% of all potential school days; range: 0–40%) were on days with reported painful episodes. There was no significant difference between the rate of missed school on the day after a painful episode (when patients might be expected to be recovering) and on other pain-free days. Of the school days missed because of painful episodes, 65% were missed when pain was managed at home, and 35% were missed when pain was managed in the hospital.

Correlational analysis showed a significant relationship between missing school on days with and on days without pain (rho = 0.523; $t = 2.37$; $df = 15$; $P < 0.05$). The average pain intensity per month was correlated with school absence on days with pain (rho = 0.599; $t = 2.99$; $df = 16$; $P < 0.01$), and with school absence on days without painful episodes (rho = 0.482; $t = 2.13$; $df = 15$; $P < 0.05$). The average analog pain score when school was missed because of pain managed at home was 55 of 100 (range: 8–86), relative to a significantly lower pain score of 30 (range: 15–47) when school was not missed. There were no relationships between the rate of school absence on days with or without pain and the percentage of days with pain, sleep quality, adherence to the diary system, or the rate of hospitalization.

Even though the children missed an average of 6–8 weeks of school within a school year, the average consecutive number of days on which school was missed was 2.7, with a range of averages of 1–9.

Discussion

Patient selection

Patterns of interaction with the health care system vary widely among patients with frequent pain and their families. Many families regularly participate in outpatient follow-up, including medical, educational, mental health, and supportive services (outpatient based management). Other families more often interact with health care professionals at the time of hospitalization (hospital-based management) (Shapiro 1993).

Families and patients who espouse outpatient-based management may have been more likely to participate in this study. We were unable to compare factors which could have influenced participation (e.g., socioeconomic status, family function, social supports, health care beliefs) between families choosing and not choosing to enroll. Additionally, the total number of patients enrolled and studied is small. Therefore, the results must be interpreted as preliminary, and not necessarily representative of all patients with frequent painful episodes. However, patients with frequent and severe pain who rely on hospital-based management are unlikely to experience less impact of the pain on activities of daily living.

Adherence

Adherence to the diary system and the study was extremely high, especially considering the nature of the task and the long-term enrollment. There were limitations, however, to determining adherence. Although we could ascertain whether the diaries were completed and returned, we do not know whether they were filled out twice a day as instructed, or whether the entire diary was filled in at one time before return. Therefore the results must be understood as an approximation of the pain experience.

Several factors may have facilitated adherence. Patients were self-selected, and their electing to participate in such a study evidenced beliefs in the value of anticipatory, non crisis-oriented intervention. The study design included methods to enhance adherence (Shapiro et al. 1990). Finally, even though we presented the diaries as a way of helping us understand the pain, many patients and families viewed the diaries as therapeutic, perceiving a benefit to themselves in recording pain and examining patterns.

We expected that pain would motivate patients to maintain participation. However, patients with more pain days had lower rates of diary completion. This result could reflect the complex inter-relationships among pain perception, coping styles, family organization, attributional style, and mood.

Home versus hospital management

The vast majority of painful episodes were managed entirely at home. Coming to the hospital for pain

management likely reflects complex interactions among such factors as pain intensity, learned behaviors, coping styles, external stresses and supports, and health care beliefs of the patient and the family.

Although the number of hospitalizations may be a sensitive indicator of illness severity for patients who tend to come to the hospital, there appears to be a group of patients for whom the rate of hospitalization does not indicate the extent and impact of the pain. If we aim to understand and reduce the impact of pain on psychosocial function and quality of life, we must include pain that is managed at home in our studies of assessment and intervention, as well as in our clinical approach to patients. The frequency of pain is an important criterion in selecting patients for treatment outcome studies. Our data suggest that in such studies the evaluation of treatment outcome may be inaccurate unless a system to track pain managed at home is included. Additionally, the fact that nearly 90% of painful episodes were managed at home in this group of patients highlights the need for programs designed to help children and families to manage their pain.

Pain patterns

The patients in this study experienced pain on a surprisingly high number of days. Clinicians often regard sickle cell-related pain as being an acute or acute recurrent pain syndrome. However, patients with frequent painful episodes are more accurately designated as having chronic or chronically recurrent pain. Since the clinical approaches to patients with chronic and acute pain differ, recognition of the pervasiveness of pain in everyday life is key.

These data were obtained during the course of an ongoing interventional study utilizing self-hypnosis and education. These interventions, which were started about 4 months into the study, may have affected the actual or reported frequency, intensity, and/or duration of pain. However, these aspects of the pain experiences are unlikely to have been increased by the interventions. Therefore, the data reported here may actually underestimate the impact of pain.

Sleep patterns

The effects of pain on sleep quality and duration were profound. Since poor or inadequate sleep may affect pain coping during the day, as well as attentiveness at school, this relationship is clinically significant. Pharmacologic and non-pharmacologic interventions targeting sleep may augment daytime performance.

Effect of pain on school attendance

This particular group of patients missed about one-fifth of their school days. Only one-half of those absences were associated with SCD-related pain. The absences on days without vaso-occlusive pain could have a number of causes, including minor infections, clinic visits, or other medical problems associated with SCD. The impact of pain and illness on psychosocial function could also affect school attendance. Families may perceive their children as vulnerable, and keep them out of school for problems that would not interfere with school attendance for most children. Additionally, falling behind in school work may develop into a pattern of school avoidance. Our finding of a relationship between the proportion of school days missed on days with and days without pain is consistent with these possibilities.

Home management of pain is often encouraged by health care professionals as a way to normalize everyday functioning. However, children with frequent pain and infrequent hospitalizations may be at significant risk of school absenteeism. Although the relationship between absenteeism and school performance has not been studied in these children, we can assume that frequent absences are likely to lead to problems. These children may be overlooked for intervention because, unlike children who are frequently hospitalized, the extent of dysfunction may not come to the attention of health care professionals.

Although these children missed an average of 6–8 weeks of school each year, they missed on average less than 3 consecutive days at a time. Thus, they do not qualify for help in states that require 1–2 weeks of consecutive missed school days for homebound tutoring, and even in states that provide more immediate homebound assistance, by the time the paperwork is filed and processed the child is ready to return to school.

Further work is necessary to delineate the impact of these absences on school performance and psychosocial status, and to develop cost effective and pragmatic ways of providing adequate educational opportunities for children and adolescents with sickle cell disease.

References

Dinges, D.F., Napping patterns and effects in human adults. In: D.F. Dinges and R.J. Broughton (Eds.), Sleep and Alertness: Chrono-

biological, Behavioral and Medical Aspects of Napping, Raven Press, New York, 1989, pp. 171–204.

Hobbs, N., Perrin, J. and Irey, H., Chronically Ill Children and Their Families, Jossey-Bass, San Francisco, CA, 1985.

Platt, O., Thorington, B.D. and Brambilla, D.J., Pain in sickle cell disease: rates and risk factors, N. Engl. J. Med., 325 (1991) 11–16.

Samuels-Reid, J. and Scott, R.B., Painful crises and menstruation in sickle cell disease, South. Med. J., 78 (1985) 384–385.

Serjeant, G.R., Sickle Cell Disease, Oxford University Press, Oxford, 1985.

Shapiro, B., Management of painful episodes in sickle cell disease. In: N.L. Schechter, C.B. Berde and M. Yaster (Eds.), Pain in Infants, Children and Adolescents, Williams and Wilkins, Baltimore, MD, 1993.

Shapiro, B., Dinges, D.F. and Orne, E.D., Recording of crisis pain in sickle cell disease. In: D. Tyler and E. Krane (Eds.), Advances in Pain Research and Therapy, Vol. 15, Raven Press, New York, 1990, pp. 313–321.

Tetrault, S.M. and Scott, R.B., Five year retrospect study on hospitalization and treatment of sickle cell anemia patient. In: Proc. 1st National Symposium on Sickle Cell Disease, Bethesda, MD, 1974.

Vichinsky, E.P., Johnson, R. and Lubin, B.H., Multidisciplinary approach to pain management in sickle cell disease, Am. J. Pediat. Hematol. Oncol., 4 (1982) 328–333.