Pain, quality of life, and coping in sickle cell disease

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Abstract

This study examined the frequency and severity of sickle related pain, its impact on quality of life, and methods of coping for 25 children with sickle cell disease, aged 6–16 years. Subjects were matched with non-affected peers and asked to complete the Central Middlesex Hospital Children's Health Diary for four weeks.

Results indicated that sickle pain occurred on average one in 14 days, and total summary pain scores indicated significantly greater pain than for controls. Children with sickle cell disease could discriminate sickle pain and did not adopt sick role responses to ordinary childhood ailments. Nearly all sickle pain was dealt with at home. Sickle pain resulted in over seven times increased risk of not attending school and was highly disruptive of social and recreational activities.

Careful assessment of sickle pain in the home environment is an essential part of a community focused pain management service, which effectively supports children's resilience and improves their quality of life.

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Sickle cell disease is a group of recessively inherited blood diseases, in which the most common type is the homozygous SS state, although the interaction of sickle haemoglobin with other globin chain abnormalities also results in clinically significant but often milder disease, for example SC disease. The patients suffer from chronic haemolytic anaemia, recurrent painful crises, and end organ failure including asplenia, stroke, avascular necrosis, chronic lung disease, and chronic renal failure. The risk of unexpected death due to infection as a result of hyposplenism, and to acute vaso-occlusive events, means that around 11% of patients do not survive to adulthood.¹ There are around four million people with sickle cell disease globally,² of whom over 6000 live in Britain and some 60 000 in North America.³

Studies of the cognitive aspects of sickle cell disease have indicated small IQ deficits, which probably indicate subtle neurological effects due to subclinical cerebral infarcts.⁴ Studies investigating aspects of psychological adjustment to the disease have tended to report an inconsistent pattern of effects including increased depression,⁵ lowered self esteem,⁶ and altered family functioning.⁴ However, although

the psychological impact of the disease should not be minimised, the current evidence also suggests high levels of successful adaptation and coping with the long term stress associated with sickle cell disease.⁷

Sickle cell related pain has been classified as an acute recurrent pain syndrome.⁸ Pain crises are difficult to anticipate and are very variable in quality, duration, location, and severity. They may occur in any part of the body, although more frequently in the extremities and may be brief (less than one hour) or last for several days. The frequency and severity of pain crises is also extremely variable, with some patients reporting no pain crises, while others report frequent pain.^{9–12}

The aim of the present study was to examine the frequency and severity of all pain and illness events experienced by children and adolescents with sickle cell disease within the home and school environment, and compare these with a control group of non-affected children; to compare the frequency and severity of pain experienced by children with SS and SC disease; to determine whether children with sickle cell disease could discriminate between 'sickle' pain and other types of childhood pain; to assess the impact of different types of pain on their quality of life, and to describe their ways of coping with the pain.

Subjects and methods

DESIGN

The study used a cross sectional case-control design using a clinic sample of children with sickle cell disease aged between 6–16 years and matched non-affected controls of similar age, sex, ethnic background, and current schooling. The children with sickle cell disease were obtained from the haemoglobinopathy register of a North London hospital (the Central Middlesex Hospital) and included both regular clinic attenders and children who attended the department infrequently. The majority of children lived in an inner city environment and control children were recruited from schools attended by the children with sickle cell disease. The control children were selected randomly by identifying children of similar age, sex, and ethnic background within the same year group as the child with sickle cell disease and then selecting the child nearest on the class register.

Ethical approval was obtained from the local ethics committee and written consent was obtained from families participating in the study.

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Table 1 Demographic details of children with sickle cell disease and controls

Measures	Sickle cell disease group (n=25)	Controls (n=25)
No (%) males	7 (28)	9 (36)
Mean (SD) age	9.83 (3.12)	10.10 (3.19)
Ethnic origin (%)		
African	15 (60)	14 (56)
Afro-Caribbean	10 (40)	11 (44)
Marital status of mother (%)		
Single	12 (48)	9 (36)
Married	13 (52)	16 (64)
No (%) of subjects with no hospital admissions in last 4 years	13 (52)	
Mean (SD) haemoglobin (g/l)		
SS	86.7 (9.3)	
SC	112.5 (10.0)	
Mean (SD) % fetal haemoglobin		
SS	9.48 (14.16)	
SC	All < 3.5	

SAMPLE

Twenty five children with sickle cell disease (14 SS and 11 SC) and 25 control children were recruited to the study. Table 1 shows the demographic and clinical data for the sample. There were no significant differences between patient and control groups for age, sex, ethnic background, and social class.

MEASURES AND PROCEDURE

Each child in the study was asked to complete the hospital Children's Health Diary (LJ Gill *et al*, unpublished) each day for a period of four weeks. The diary followed a semistructured format in which children recorded information about their general health and wellbeing using a five point rating scale and recorded the presence of 24 common childhood symptoms (for example, cough/cold, tiredness, headache). Detailed information on each specific pain event was recorded including its intensity, location and duration, its impact on nine different types of activity (going to school, doing school work, doing sport, sleeping, eating and drinking, watching television, going out, seeing friends, taking part in favourite activities), and the way of coping adopted by the child and family (taking medication, going to see a doctor, going to bed, talking to parents, teachers, and other relatives). In addition, the children with sickle cell disease were asked whether their pain was related to the disease or not.

STATISTICAL ANALYSIS

A total of 1400 recorded diary days were coded and exploratory analysis was carried out by drawing up descriptive tables treating days as cases. In order to ensure independence of observations, statistical comparisons between groups were carried out using summary scores for each subject. For example, a total summary pain score for each subject was computed by summing the combined intensity and duration scores for each pain event.

Children with sickle cell disease were asked to rate whether they perceived the pain to be 'sickle related', or not. Mean pain, impact, and coping scores were computed for three types of pain: sickle pain, non-sickle pain, and controls' pain. Statistical comparison between these groups was carried out using one way analysis

of variance (ANOVA) with *a posteriori* contrasts of all three groups using a modified LSD (Bonferroni) test with significance level at 0.05. In order to take into account within subject variation (for which the ANOVA does not control), additional comparisons were made between subjects with both sickle and non-sickle pain events, using paired significance tests. Other comparisons between children with sickle cell disease and controls used parametric or non-parametric tests depending on the type of distribution of specific variables. In general, more conservative non-parametric tests (Mann-Whitney U test) were preferred as the distribution of some outcome variables was not normal. One tailed tests of significance were used when the direction of effects was clearly predicted prior to data collection.

Results

GENERAL HEALTH

Results from daily ratings of 1400 diary days did not indicate significant differences between children with sickle cell disease and controls on 'quality of day' ratings and general health symptoms. Children with the disease rated their health as 'poor' for 4% of days compared with 2% for controls. However, control children tended to have more positive ratings ('very good' days) compared with children with sickle cell disease (46% *v* 25% respectively). From the health symptom checklist (24 items) children with sickle cell disease reported significantly more frequent symptoms only with respect to 'waking during the night' (Mann-Whitney U test, $p < 0.009$). Children with sickle cell disease reported this problem nearly five times more frequently (19.7% *v* 4.6% nights) than controls. None of the children with sickle cell disease reported regular bed wetting problems (four reported a single wet night). Children with sickle cell disease did not have significantly more days with symptoms (55.6% *v* 48.1% days) than controls.

PAIN EVENTS

Children with sickle cell disease reported 240 days with a pain event during the 700 diary days (35%) compared with 182 days with a pain event (26%) reported by controls. The mean total number of days (out of 28 days) in pain per subject did not differ significantly between children with sickle cell disease and controls (mean (SD) 9.60 (7.59) *v* 7.28 (5.97) days). The two groups did not differ with respect to mean intensity scores (38.92 (16.23) *v* 35.91 (20.58)) but children with sickle cell disease had pain for a significantly longer duration compared with controls (42.27 (26.22) *v* 26.67 (22.59); Mann-Whitney U test, $p=0.019$). This increased frequency and longer duration of pain for children with sickle cell disease was reflected in their higher total summary pain scores (2.42 (3.05) *v* 1.14 (1.83); Mann-Whitney U test, $p=0.036$). However, 80% of children with a total summary pain score of greater than 3.0 were from the sickle cell disease group (fig 1), including three children with SC disease. Overall children with

Table 2 Comparison of sickle pain, non-sickle pain, and controls' pain

	Sickle cell disease group (n=25)		
	Sickle pain	Non-sickle pain	Controls (n=25)
Severity			
Mean summary pain score (0-1)	0.27	0.18	0.12
Mean weighted pain duration score 0-100 (100 = all day continuously)	55.58	36.99	26.67
Impact (proportion of days with pain by impact)			
Did not go to school	0.36*	0.04	0.02
School work not completed	0.30*	0.03	0.03
Missed sport	0.64*	0.08	0.07
Missed favourite activities	0.62*	0.06	0.03
Coping (proportion of days with pain by coping response)			
Took medicine for pain	0.54*	0.09	0.06
Went to bed due to pain	0.32*	0.03	0.02

* Difference between sickle pain and non-sickle pain was significant at the 0.05 level.

NB. All differences between sickle pain and control pain were significant at the 0.05 level.

sickle cell disease experienced more than twice as much pain as their non-affected peers. This difference in pain was not generalised to all types of pain experience. Children with the disease did not have higher rates of headaches, chest pain, back pain, or abdominal pain but did report significantly higher rates of limb pain compared with controls (Mann-Whitney U test, $p < 0.022$).

DISCRIMINATING BETWEEN SICKLE AND NON-SICKLE PAIN

For children with sickle cell disease the minority of pain events were perceived as not being related to the disease (27%). Out of 25 children with sickle cell disease, 13 children perceived none of their pain experience as being related to the disease. Nineteen separate sickle pain crises were identified from the 65 sickle pain event days. Twelve crises lasted for less than two days, one for three days, and six for 7-9 days. Only one of these crises resulted in attendance at hospital.

For the children with sickle cell disease 175 days with a non-sickle pain compared with 182 days with a pain reported by controls. The proportional increase in pain events between those with sickle cell disease and controls is accurately accounted for by the sickle related pains (fig 2). The combination of higher intensity and longer duration resulted in sickle related

pains being significantly more severe than pain events reported by controls (table 2).

Approximately 30% of sickle related pain was attributed to preceding events such as cold weather (14%), exercise (12%), and coughs (3%). Additionally, for one child, sickle related pain was clearly triggered by physical trauma such as falling over. There was an increased frequency of reported symptoms on the day before the beginning of a pain crisis. Altogether 75% of pain crises were preceded by a symptom indicating general poor health (for example, tiredness, cough/cold). There was also an increased frequency of particularly disturbed sleep (43%).

IMPACT OF SICKLE CELL DISEASE ON QUALITY OF LIFE

For all nine everyday activities recorded in the diary, the impact of pain events on everyday activity for children with sickle cell disease was much greater than for controls (table 3). Children with sickle cell disease missed nearly 2% of school days due to pain events and were seven times more likely to miss school due to pain than controls. Similarly high rates of disruption were evident for being unable to take part in their favourite activities, doing sport, or seeing their friends. One child was unable to do sport for 26 out of 28 days. Children with the disease had three times more

Figure 1 Total summary pain scores for children with sickle cell disease (sickle cell anaemia (SS) and SC disease (SC) and controls (over 28 days).

Figure 2 Mean duration of pain for the total number of days with sickle pain, non-sickle pain, and controls' pain.

Table 3 Comparison of children with sickle cell disease and controls: mean (SD) and median (range) number of days when pain prevented an everyday activity, over a 28 day period

Activity	Sickle cell disease group (n=25)	Controls (n=25)	Relative risk
Going to school	1.84 (3.45) 0.00 (0-12)	0.24 (0.66) * 0.00 (0-3)	7.67
Doing school work	1.32 (2.87) 0.00 (0-12)	0.40 (1.08) 0.00 (0-5)	3.30
Doing sports	3.24 (5.95) 1.00 (0-26)	1.68 (3.30) 1.00 (0-16)	1.93
Eating and drinking	0.96 (1.43) 0.00 (0-5)	0.44 (0.92) 0.00 (0-4)	2.18
Watching television	0.32 (0.56) 0.00 (0-2)	0.16 (0.47) 0.00 (0-2)	2.00
Going out	2.60 (5.02) 0.00 (0-21)	0.48 (0.87) 0.00 (0-3)	5.42
Seeing friends	1.08 (1.66) 0.00 (0-6)	0.16 (0.37) * 0.00 (0-1)	6.75
Taking part in favourite activities	2.68 (5.91) 0.00 (0-27)	0.40 (0.71) 0.00 (0-3)	6.70
Total No of impacts (due to pain) over 28 days (0-252)	15.71 (22.79) 4.00 (0-75)	4.48 (6.27) 2.00 (0-23)	

* $p < 0.05$.

'impacts' than controls (15.7 *v* 4.5). As with pain scores, six out of seven children with scores greater than 20 were from the sickle cell disease group. The impact of a pain event was much greater when the event was perceived as being related to sickle cell disease (table 2). Nearly one third of sickle related pain days resulted in children not attending school whereas non-sickle related pain events rarely resulted in non-attendance at school. There was no difference between the impact of non-sickle pain for children with sickle cell disease and controls.

COPING WITH PAIN

For both groups, the most common response to coping with pain was to talk to a parent, and there was no difference in the frequency of discussing pain events with either siblings, friends, or teachers. For sickle related pain children with sickle cell disease were more likely to take medication (54% of pain days) than for non-sickle pain (9%) or compared with controls (6%) (table 2). The main medication used by both groups was paracetamol (69% of all medication used). Four children with sickle cell disease used codeine based compounds and one SS patient was admitted and opiate medication was used. For 32% of sickle pain days, children with sickle cell disease stayed in bed all day whereas this was a rare event for controls (2%).

Finally, children with both sickle and non-sickle pains (n=11) were compared on all the pain severity, impact, and coping variables (listed in table 2, ANOVA), using paired *t* tests. The results showed that all the differences between sickle and non-sickle pain events that were significant on the ANOVA (table 2), were again significant at the 0.05 level, using paired *t* tests.

Discussion

Children with sickle cell disease had similar amounts of minor ill health (excluding sickle cell disease pain) and non-sickle pains (for example headaches) to their peers, and coped in a similar way. Thus there was no evidence that children with sickle cell disease tended to

put themselves in a sick role by interpreting minor health events (for example colds, headaches, etc) in a more anxious and/or sensitised way than control children. Also, they were able to discriminate accurately between sickle related pain and other types of pain. Pains perceived to be due to sickle cell disease were significantly different from non-sickle pain in terms of location, duration, intensity, and overall severity.

Sickle pain only once resulted in hospital admission despite it occurring on average one in 14 days. This estimate appears to be lower than the previously reported frequency of one in four days,¹¹ but this difference was probably due to sampling differences. The Shapiro study only included subjects with at least three vaso-occlusive crises over the previous year. According to Platt *et al* this would include only the most severe 5% of the sickle cell disease population.⁹ In our own sample, 48% had not had a vaso-occlusive crisis requiring hospital admission during the previous four years. Examination of the more severe children in our sample would broadly support Shapiro's much higher estimate.

CLINICAL IMPLICATIONS

Firstly, sickle pain, coped with at home, is a common experience for a large proportion of children with sickle cell disease and is likely to be under-reported in routine outpatient follow up appointments. The children in this study reported far fewer sickle pain events when attending the paediatric clinic and tended to minimise the impact of the disease on their daily lives. Children may be apprehensive about talking about their pain and/or other problems as this may be seen to increase the possibility of further blood tests or even a hospital admission.

Secondly, clinical management of sickle cell disease needs to take account of its variability, differences in coping styles, and the range of impact on daily life. Such variability indicates the importance of collaborative assessment involving the parent, child and clinician, before individually tailored advice is given. As nearly all sickle pain is dealt with at home, systematic

assessment of current pain and coping methods needs to be focused on supporting successful pain management in the home environment. This can be done in two ways. Firstly, retrospective accounts of health events such as pain have very limited reliability.¹¹ The use of a health diary for a brief period (2–4 weeks) may be extremely helpful in establishing the pattern, severity, and impact of sickle cell disease on the child's life. Secondly, the clinical team need to discuss with parents and children the ways that they have successfully coped with pain at home. Consultation can therefore become a process of supporting resilience as well as identifying clinical problems.

The identification of reliable triggers for sickle pain remains elusive. However physical trauma clearly triggered sickle pain for one child. Reported symptoms before onset of a sickle related pain (for example tiredness, cough/colds, and dehydration) were generally quite diverse, and would be difficult to use as predictors of a developing pain crisis. It is probable that minor ill health contributes to the risk of a pain crisis. It remains unknown whether psychological factors also play a part in increasing risk of sickle pain. More work is needed to develop a multivariate model that would have useful predictive power to enable patients to anticipate the beginning of sickle cell disease pain events.

Despite the resilience of children with sickle cell disease, this chronic disorder has a significant impact on the physical, psychological, educational, recreational, and social life of these children. A comprehensive health care

service that aims to lessen this impact needs to be a genuinely multidisciplinary service that facilitates effective cooperation between health, education, and social agencies. However familiar this assertion may be, the provision of such services for many children with sickle cell disease still remains to be established.

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