ORIGINAL ARTICLE

Impact of Undertreated Sickle Cell Pain in the Caribbean

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ABSTRACT

Objective: *Undertreated pain around the world includes the acute and chronic pain caused by sickle cell disease (SCD). In collaboration with a Caribbean association that aims to provide assistance to those diagnosed with SCD, we surveyed adults with SCD about pain management and impact of SCD pain.*

Methods: *Participants were recruited from a group of 55 adults with SCD. A survey was administered to those who agreed to participate. Questions centred on their self-assessed level of pain due to SCD, the extent to which that pain interferes with daily activities, and how they seek and obtain pain relief.*

Results: *Responses were received from 39 participants (female: n = 28, 72%, male: n = 11, 28%; mean age: 31.6 (SD ± 13.7) years. Sickle cell disease pain significantly disrupts participants' daily activities (62%), mood (72%), work (64%) and sleep (69%). Prescription medicine was ineffective for 41% and about half (n = 19) sought alternate means of relief.*

Conclusion: *Sickle cell disease pain is undertreated in the Caribbean, disrupts daily activities and affects quality of life by impinging on education, employment and marital status. Sickle cell disease and other types of pain can be clinically managed safely, effectively and inexpensively. By failing to palliate and overcome the problem of undertreated pain, healthcare systems and providers contribute to socio-economic amongst other repercussions for sufferers, their families and caregivers, and their nations.*

Keywords: Caribbean, sickle cell disease, undertreated pain

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INTRODUCTION

At some point in time everyone experiences chronic or acute pain. Socio-economic, cultural and other factors hinder access to effective pain relief and there are ethical implications (1–3). Among other things, the inability to obtain relief makes patients more vulnerable to depression and anxiety, and undermines doctor-patient relationships and public trust in medicine; this constitutes an injustice (4–6). Withholding pain medications or treatments leads to unnecessary suffering and violates human rights, but billions of patients around the world are deprived of effective relief because doctors' knowledge of pain management is deficient, and because legal barriers hinder the prescription of and access to essential pain medications (7). Undertreated pain is a global problem with emotional and economic costs for individuals, populations and health systems.

One source of chronic and acute pain in adults and children is sickle cell disease (SCD), an inherited blood disorder with increasing global prevalence (8, 9). The prevalence, severity and impact of SCD pain are widely underestimated and undertreated but stem from vaso-occlusive episodes (10–12). Sickle cell disease pain accounts for most SCD hospital admissions and the average stay is seven to eleven days (13). Sickle cell disease pain is severe and frequent enough to impede daily activities including school attendance (13, 14). Undertreated pain from SCD or any source typically generates repeated efforts to obtain relief but such behaviours are perceived as 'drug-seeking' and deter doctors from prescribing effective pain relief (11). This sows distrust between doctor and patient, and is worsened by the lack of sympathy and preoccupation with drug addiction demonstrated by many hospital staff (6).

Genes that contribute to SCD are most frequently found in those of African descent and are therefore common in Caribbean populations (15). Most English-speaking Caribbean nations offer at least a minimal level of universal

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healthcare and have rising socio-economic and health indicators but, as in other developing nations, their SCD prevalence is not known and there is little medical or social services support to manage or control SCD pain (9).

We investigated how often and severely patients in the Caribbean experienced SCD pain and to what extent this interfered with their daily life. The study was designed in collaboration with the Sickle Cell Association of Grenada (SCAG), a nonprofit organization that aims to support the needs of sickle cell patients and their families in Grenada. It constitutes an unofficial registry for SCD since local resources limit the capacity to establish or maintain registries for any condition, which is the case in English-speaking Caribbean nations with similar socio-economic and health indicators. The first documented patient with SCD was a Grenadian who had been enrolled in dental school in 1904 in the United States of America [USA] (16). We hypothesized that SCD pain in the Caribbean is undertreated and impacts negatively on education, employment, marital status and enjoyment of life.

SUBJECTS AND METHODS

This study was described to and discussed with members of SCAG during a general meeting. The recruitment population included SCAG's registry of 55 adults with SCD aged 18 years and over. Individual invitations to participate were subsequently made by phone to each member who met the recruitment criteria. Three attempts were made to contact each potential participant. Participants were given the choice of completing the survey in a phone interview, in person, or independently in writing. The study received approval from the Institutional Review Board at St George's University in Grenada.

Participants were surveyed using a modified version of the 'Brief Pain Inventory' (17). Some questions were omitted and others altered for relevance to the study goals and the host population. Using a 10-point scale where 'one' designates 'no interference' and '10' designates 'interferes completely', participants were asked to assess the impact of SCD pain on their mood, ability to walk, work and sleep, relations with other people and enjoyment of life. A score of eight or more was categorized as 'severe'. Other questions addressed demographics, how often within the past year the participant had sought medical care for SCD pain and how many different doctors they had seen for it, and the effectiveness of prescribed pain medications.

Mean and standard deviations (SD) were reported for continuous variables while frequencies and proportions were reported for categorical variables. Associations between demographic characteristics (gender, age, marital, educational and employment status) and severity of interference in general activity were investigated. Odds ratios (OR) with corresponding 95% confidence intervals (CI) were calculated to determine the strength of association with each demographic characteristic. All analyses were completed using StatPlus:mac LE, version 5.9.2.0 (AnalystSoft Inc.).

RESULTS

Of the 55 adults registered with SCAG, 39 (71%) participated. All participants (n = 39) were of Afro-Caribbean descent. Sixteen could not be contacted, were deceased, or declined to participate. Participants had a mean age of 31.6 ± 13.7 years and were predominantly female (n = 28; 72%). Most were unmarried (n = 30; 77%) and unemployed (n = 27; 73%). Less than half had completed secondary school (n = 17; 46%).

Twenty-five participants (65%) had seen more than one doctor for their sickle cell pain within the previous year. Nineteen had visited the hospital at least three times for SCD pain within the previous year, and the same number had experienced severe pain in the week prior to being surveyed. Most reported that SCD pain severely interferes with their general activity (62%), mood (72%), ability to walk (59%), work (64%) and sleep (69%). Sickle cell disease pain also causes moderate to severe interference in their relations with other people (52%) and their enjoyment of life [74%] (Table 1).

Participants were asked which of the following strategies they had used to relieve their SCD pain: prescription medicine, rest, traditional 'bush' medicine, application of heat, and 'other' (Table 2). About half (n = 19) reported using strategies other than prescribed medications and 44%

Table 1: Self-reported pain and its impact on quality of life among sickle cell patients in Grenada (n = 39)

"Fill in the one number that describes how, in the past year, pain has interfered with your":	Little to no interference (score 1–3) n (%)	Moderate interference (score 4–7) n (%)	Severe interference (score 8–10) n (%)
General activity	2 (5%)	13 (33%)	24 (62%)
Mood	1 (3%)	10 (26%)	28 (72%)
Walking ability	4 (10%)	12 (31%)	23 (59%)
Work (at home or in workplace)	5 (13%)	9 (23%)	25 (64%)
Relations with other people	19 (49%)	8 (21%)	12 (31%)
Sleep	8 (21%)	4 (10%)	27 (69%)
Enjoyment of life	10 (26%)	11 (28%)	18 (46%)

Table 2: Efficacy of prescribed medications (n = 39)

	"When you take medicine for sickle cell pain, how many hours does it usually take before the dosage takes effect?" n (%)	"When you take medicine for sickle cell pain, how many hours does it usually last before the pain returns?" n (%)
Pain medication does not help at all	17 (44%)	17 (44%)
Up to or less than 1 hour	6 (15%)	1 (3%)
1–4 hours	10 (26%)	14 (36%)
4–12 hours	1 (3%)	1 (3%)
12 hours or more	0 (0%)	1 (3%)
Does not take pain medication	5 (13%)	5 (13%)

(n = 17) said that medication helped. When asked what exacerbated their sickle cell pain, 51% (n = 20) responded that physical activity did so and 10% (n = 4) stated that side effects from the medication worsened their pain. Of the 17 participants who reported using prescription medication, 10 (59%) reported that it took up to four hours for the pain relief to take effect and 14 (82%) reported that the duration of the effect was only between one and four hours.

Unemployed participants had an eight-fold increased odds (OR: 8.4; 95%CI: 1.4, 52.8) of severe interference of daily activities from SCD pain compared to those who were employed. While the odds ratios of age (OR: 2.1; 95%CI: 0.4, 11.9) and marital status (OR: 2.9; 95%CI: 0.3, 26.7) were elevated, no statistically significant association with severe interference was detected for age, gender, marital status, education, or reliance on prescription medication.

DISCUSSION

Grenada has no data on the prevalence of SCD or SCD pain in its population of about 100 000 and SCAG was the only source through which we could identify SCD patients. The willingness of 71% of its small membership to participate is suggestive of their need for more attention and support. That most participants were female corresponds with the broad tendency toward greater use of medical and social support by women than men. The choice of most participants to complete the survey by phone is consistent with Grenada's cultural preference for verbal rather than written communication.

The lack of phone service among about 20% of potential participants suggests that they have a lower socioeconomic status than other participants and it may be a potential source of bias. The study did not investigate co-morbidities associated with SCD or SCD pain. Limitations include potential recall bias in participant self-reports and the small sample size. Participation was elicited from all parishes of Grenada, and variability in data collection was controlled by using only one investigator to administer the survey.

Impact of sickle cell disease pain

The negative impact of SCD pain on participants is evident in its moderate to severe interference with their enjoyment of life (74%); most were unmarried (77%), unemployed (73%) and did not complete secondary education (54%). The odds of reporting a disruption of daily activities because of pain was eight times higher among those who were unemployed compared to those who were employed. Combined with evidence that SCD pain significantly impedes school attendance, we infer that the disruption of a participant's education by SCD pain was severe enough to diminish his or her ability to gain employment (14, 18). The respective odds ratios for marital status and education are not statistically significant but it is plausible that, as children, participants' SCD pain hindered their social activities enough to impede their development of interpersonal skills essential to marriage and employment. Providing aggressive pain relief improves outcomes and productivity, so its provision to children with SCD pain would increase the ability to develop healthy relationships, obtain education and become productive adults (5).

More than half of the participants described their SCD pain as severe during the week prior to being surveyed, and more than half saw two or more doctors and made three or more hospital visits for their SCD pain in the year prior to being surveyed. Relatively few obtained relief from prescription medication and this may reflect, among other things, the Caribbean's reliance on pethidine as the standard of care instead of oral morphine which is globally recommended for severe or moderate pain (1–3, 19). Failing to provide immediate release oral morphine to all who rate their pain as moderate or severe violates the right to the highest attainable standard of health; substituting sustained release and/or injectable formulations for oral morphine is poor clinical and public health practice and governments, pharmaceutical companies and healthcare institutions have a responsibility to ensure accessibility of oral morphine at a reasonable cost (20).

Undertreated pain is a global problem that contributes to poor clinical outcomes and co-morbidity for depression (1,

2, 5, 19). It stems from misconceptions about pain, patient truthfulness, substance abuse, opioid medications and overly stringent regulations on the medical use of opioids (2, 6, 12, 19). There are relatively few pain or palliative specialists anywhere, and palliative care is usually delivered as end of life care so it is largely unavailable to those with SCD and non-terminal conditions (2). This is compounded by the greater allocation of resources to prevention and cure rather than to relief of pain and symptoms, and by the inadequate exposure of doctors to clinical guidelines for managing pain (1, 2). Other countries with similar sickle health burdens attribute poor management of the disease to poor research, management techniques and stigmatization (21, 22).

The frequency of acute episodes of SCD pain also correlates with early death of adults (8). Providing effective relief would likely alter their prognosis and increase their ability to contribute economically and in other ways to their families, communities and nations. Efforts to frame pain and symptom relief as essential to improving outcomes, like recovery and survival rates, would help to integrate effective pain relief into general practice. Exploration of beliefs, policies and health outcomes in varied contexts is needed to provide insight into how to combat the global scourge of undertreated pain.

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