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ORIGINAL RESEARCH

Patient and Caregiver Perspectives on Care-Seeking During a Vaso-Occlusive Crisis in Sickle Cell Disease: Results from Qualitative Interviews in Canada

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Purpose: There is little research on care-seeking preferences during active pain crises for sickle cell disease (SCD) patients and their caregivers. The objective of this study was to identify relevant, patient or caregiver narratives of the pain crisis experience, to understand the factors that contribute to care-seeking during a pain crisis, and to identify preferences when making care-seeking decisions during a pain crisis.

Patients and Methods: Qualitative semi-structured interviews were conducted with Canadian residents with a self-reported SCD diagnosis, who were either ≥18 years of age or an adolescent between the ages of 12–18. Interviews were hosted virtually, audio-recorded, and transcribed verbatim.

Results: A total of 23 individuals participated (74% female; 26% male), including six adolescents with parent dyads and 11 adults. Almost all (N = 21, 91.3%) participants were Black/African American. Participants mentioned many factors that influenced care-seeking decisions, mainly the symptom and pain experience; institutional factors (waiting time, the presence of and adherence to treatment guidelines, and the empathy or racial bias felt from medical staff); and subject-level factors (age and a flexibility in daily responsibilities).

Conclusion: This study identified important institutional and subject-level considerations involved in care-seeking decisions. Most importantly, this study highlights the racial stigma faced by many patients when care-seeking in the ER and the lack of care protocol implemented, which hinders care-seeking in a dedicated medical facility. From the patient perspective, these are clear gaps to fill to encourage patients to seek and receive the care they deserve.

Keywords: vaso-occlusive crisis, sickle cell disease, pain crisis, care-seeking, standard of care

Introduction

Sickle cell disease (SCD) is a hereditary disease, originating from a single point amino acid mutation of the hemoglobin gene, modifying the shape of red blood cells, and leading to reduced or blocked blood flow.[1,2] This rare blood disorder affects approximately ~100,000 people in the United States and approximately ~5000 people in Canada.[3,4] The most common manifestation of SCD is vaso-occlusive crises (VOC), when pain is caused by obstructed microcirculation from the sickled red blood cells.[5] These acute episodes, or pain crises, can present suddenly but can also present gradually over a period of time. Patients describe SCD pain in a myriad of ways, suggesting SCD pain is complex and multifaceted in etiology, encompassing ischemic, inflammatory, and neuropathic components.[6] Common health problems related to SCD include severe pain in the joints, chest, abdomen, back, as well as progressive organ damage, infection, anaemia, and stroke.[7]

Some patients experiencing a pain crisis may attempt to manage their crisis at home either before or instead of seeking treatment in dedicated medical facilities. Despite free access to care in Canada, the unwillingness of some

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patients to seek care in dedicated medical facilities may result from previous experiences with inconsistent care-seeking protocols and evidence-based treatment guidelines for patients with SCD, and a lack of understanding from medical faculty.[8] Notably, the consequences of not care-seeking in the appropriate setting include prolonged crises and even morbidity.[9] Despite studies noting factors other than medication can influence care-seeking, such as cultural, social and demographic factors, cost, social networks and biological signs and symptoms,[10] little is known about the patient and caregiver preferences for care, and the specific factors that drive specific care-seeking decisions during pain crises. Qualitative studies in particular can provide detailed insights into the symptom experience and the understanding of care-seeking behaviour and preferences for pain management.

The objective of this study was to identify relevant, patient or caregiver narratives of the pain crisis experience; to understand the factors that contribute to care-seeking during a pain crisis; and to identify patients and caregiver's attitudes and preferences when making decisions regarding care-seeking during a pain crisis (at home or a dedicated medical facility).

Materials and Methods

Study Population

English or French speaking patients older than 18 years old and residing in Canada, with a self-reported physician diagnosis of SCD, were recruited to participate in a qualitative interview study. For SCD patients between the ages of 12–18, we relied on dyad reporting from both the caregiver's and adolescent's perspective. Adults and adolescents with SCD who had experienced 10 pain crises or more since their diagnosis and at least one pain crisis in the previous six months were eligible for the study. Adult or adolescent patients were ineligible if they also had another unrelated condition or chronic illness which made it difficult to evaluate treatment decisions.

Eleven adult patients and six dyads of caregivers and adolescent patients were recruited, totalling 23 study participants.

Participants were recruited in collaboration with the Sickle Cell Disease Association of Canada/Association d'anémie falciforme du Canada (SCDAC/AAFC), the Association d'anémie falciforme du Québec, and a market research agency, Global Perspectives. Methods of enrolment comprised of established channels, including the proprietary patient database, consumer recruiter networks, medical recruiter networks, support groups and nurses, as well as patient key opinion leaders and social media. The patient groups further supported the recruitment process by direct contact through their contact list. Eligibility was assessed using a study screener, at which sociodemographic and information about disease severity was also collected. Severity was not reported by adolescent patients in a dyad, but rather by their caregiver.

Interview Conduct

A targeted literature review and consultation with clinical experts was conducted to gather insights on common symptoms of SCD, manifestation and impact of pain crisis on the patient, current treatment practices, and perceived factors that contribute to care-seeking habits. The data obtained was used to inform the development of two semi-structured interview guides for use in the concept elicitation qualitative interviews, one interview guide for patients 12 years and older, and one interview guide for caregivers of children between the ages of 12–18. Questions in the interview guide covered symptoms, HRQL, current treatment, factors contributing to care seeking during a pain crisis, and treatment preferences. Adult patients over 18 years and adolescent patients enrolled in a dyad were both interviewed using the same patient interview guide.

One-hour telephone interviews were conducted with each participant, moderated by trained interviewers following a semi-structured interview guide. Verbal informed consent was collected for each participant prior to the telephone interviews; adolescent participants provided assent, and their caregiver provided consent. Interviews were audio-recorded, then transcribed verbatim and anonymised prior to data analysis; additionally, French transcripts were forward and back translated to US English. Interviews took place between December 2020 and March 2021. The study was approved by a central institutional review board, Salus IRB.

Data Analysis

Each transcript was checked for quality and coded using a codebook, in which codes were grouped under themes generated from topics in the interview guide. Initial codes were based on pre-defined concepts and themes; analysts added data-driven codes that emerged during the interviews. Qualitative thematic analysis of the interview data was performed by the one analyst using a specialist software (MAXQDA 2020). The lead analyst reviewed several transcripts to develop a preliminary codebook. Before the codebook was finalized, a second analyst familiar with the dataset reviewed the codebook for quality and consistency. Descriptive statistics was used to analyse the collected socio-demographic data.

Results

In describing results, we use the terms "caregivers" to refer to caregiver participants; the terms "adolescent patients", to refer to children in a dyad, and "adult patients" to refer to adult patients not in a dyad; and the term "participants" to refer to all types of participants in the study.

Participant Demographics

The demographics, clinical characteristics, and current medication of participants who took part in the interviews (N = 23) are presented in Supplementary Data Tables 1 and 2. There were predominately more female interview participants compared to males (74% and 26%, respectively); and close to all (N = 21, 91.3%) participants characterized themselves as Black/African American. Regarding education, all but two adult patients (N = 9) held either a Bachelor's, or an advanced degree at the time of the interview. Education level within caregivers varied, with four (67%) caregivers having completed some college, and two (33%) caregivers receiving a Bachelor's degree. Adult and adolescent patients were on a broad range of treatments at the time of the interview; the three most common were hydroxyurea (N = 9, 39%), folic acid (N = 10, 43%), and rescue treatments (N = 8, 35%), which were treatments taken as needed during pain crises. Of note, an additional three patients took hydroxyurea in the past but were no longer taking the treatment at the time of the interview.

Many patients who participated in the interviews had been diagnosed with SCD at birth and had experienced a multitude of pain crises over the years, with some patients experiencing pain crises multiple times per year. Adult patients experienced an average of four crises over the previous six months from screening. As described by their caregiver, adolescent patients experienced an average of 10.5 crises over the previous six months from screening. All patients enrolled in our study ranked their SCD as either moderate (N = 10), severe (N = 9), or very severe (N = 4).

Pain Crisis Experience

Symptoms During Pain Crises

While all participants noted experiencing pain, all but three participants (N = 20, 87%) noted feeling pain very early on in a crisis, occurring in various locations on the body. Common areas for pain were the back, arms, legs, and chest, with a few patients noting pain also starting in their hands. Other symptoms noticed early in the pain crisis included manifestations of anemia, such as jaundice (N = 6, 26%) and/or fatigue (N = 4, 17%), often occurring before any other symptoms.

Given the variability in pain crisis experiences between patients, a wide variety of other symptoms mentioned as pain crises progressed, are listed in the Supplementary Data Table 3. As expected, pain was the most consistent symptom noted by all participants, followed by fever, dehydration, changes in internal body temperature, swollen or discoloured eyes, headache, vomiting, and fatigue.

Impacts on Quality of Life

Patients noted a variety of impacts on their daily lives when they experienced a pain crisis. The presence of these impacts had the potential to influence attitudes and beliefs for care-seeking at the time of the crisis, and also when making decisions in the future about where to seek care. Data on caregiver impacts did not influence care-seeking decisions and is therefore, not presented in this manuscript.

School/Work

Many participants who were attending school or held a permanent job at the time of the interview mentioned difficulties with performance and attendance. Often times, if a patient was experiencing a pain crisis, they would self-care at home or seek care in a dedicated medical facility if unable to manage at home, both leading to reduced attendance at school or work. Also, long waiting times at a dedicated medical facility were described as having a great impact on the academic and work functioning for patients with SCD.

Social Functioning

A variety of social implications were described such as the inability to spend time with family and friends, and partake in hobbies. It was common to see patients adjusting their lifestyle in order to accommodate the possibility of a pain crisis, such as taking part in activities that require minimal physical effort.

Mental/Emotional Impacts

The emotional impacts of experiencing pain crises included anxiety, hurt feelings, loss of self-esteem, and frustration/anger. Some feelings of sadness and depression were noted, due to the inability to perform physical activities and the impact on their social life. Other emotional impacts mentioned included an eagerness or irritation for the pain to subside, disappointment for cancelled plans, self-consciousness for performing tasks slower than others, fear of health-care provider's stigma and lastly, fear of complications as crises progress.

Treatment Experience

Often times, patients were able to feel the onset of a pain crisis, pre-emptively taking pain medications at home to combat the crisis. When medications were non-efficacious alongside other relief strategies, most participants reported that they would consider care-seeking in a dedicated medical facility, where they could receive further intervention such as oxygen, intravenous fluids, and morphine. Notably, very few of the adult patients (2/11, 18.2%) shared it was very likely they would visit a dedicated medical facility in the event of a pain crisis.

Pain Crisis Management Strategies

Patients shared that they have learned to apply various management strategies to provide relief to, or delay the onset of, a pain crisis. Caregivers were slightly more likely to administer medication early on in a crisis, or after certain triggers were realized. Among the six dyads, four (67%) caregivers mentioned proactively giving medication to their child while their pain was mild, a strategy not mentioned by their respective child. Alternatively, six of the 11 adult patients (55%) shared they proactively took medication with mild pain symptoms, while the remaining adult patients first implored strategies such as massaging the area or staying hydrated instead of medication. Interestingly, of patients who were asked (N = 13), no meaningful instruction on how to manage a crisis and when to seek help was provided by their haematologist; three patients (23%) did not answer the question when asked, eight patients (62%) were not given instruction on how to manage a crisis, and two patients (15%) shared instruction was given; however, they did not find it meaningful. It is notable that these strategies were sought either before or instead of seeking care in a dedicated medical facility.

Preferences for Care-Seeking

Participants mentioned a variety of factors influencing their care-seeking behaviours; some factors that pushed patients into their preferred medical facility and some that kept patients at home. Outlined below are a number of factors influence care-seeking behaviours, such as the symptom experience, hospital characteristics, and subject-level factors.

Symptom Experience Factors

Many of the factors to seek care at a dedicated medical facility included the symptom experience, such as back pain, headache, acute chest syndrome or difficulty breathing, dizziness, and fever. A general intensity of all symptoms experienced or a persistent high pain level (N = 20, 87%) was a contributor to care-seeking at a dedicated medical facility.

Institutional Factors

There were also a few factors that influenced the decision to seek care at a dedicated medical facility, such as the presence of machines to monitor patients, the fact that patients could feel some relief with the administration of morphine or an IV, and the presence of psychiatrists or social workers that can support patients (N = 10, 43%). Other positive institutional aspects include the familiarity of the medical facility and the implementation of a protocol specific (N = 11, 48%) to how a pain crisis should be treated upon admission to the emergency room (ER). One patient mentioned,

> There's a really great hemoglobinopathy program that's set up, so that at least the basic things — [hydration], we need to get treatment ASAP, no long waits in the wait[ing] room … oxygen and IV … If all of these things are dealt with early, then the patient can get out early, and it doesn't get worse. [CAD-104-P]

While participants understand the need to seek care outside of the home, there are some institutional factors that hinder patients from going to the ER. Unfortunately, not every patient has a protocol in place at their preferred medical facility. For these patients, the lack of a protocol in place at their preferred medical facility is a factor that motivates them to treat their pain crisis at home. Interestingly, one patient noted the over-implementation of this pain crisis protocol at their preferred medical facility, where even for pain unrelated to a VOC, the ER staff could only attribute VOC to be the cause of the pain. This adolescent patient shared,

> There was one time where the doctors misdiagnosed me, because they thought — well, it was one doctor. I was having pains, and he thought it was sickle cell pain, so he didn't think it was serious. But then my parents made me go to another doctor, and it turned out I had gallstones, but the doctor kept saying it was just sickle cell pain, so I had to go get surgery [CAD-211-P]

Participants mentioned specific hospital characteristics (N = 13, 57%) such as waiting time, comfort level, the smell or sounds of a hospital, previous experiences such as the inability for nurses to find their veins, and the inability to have childcare for their children if admitted to the hospital that prevented them from care-seeking in the hospital. Pain crises for these patients last significantly longer due to the prolonged management of pain at home, and the delay in treatment if care is eventually sought at the hospital. Some patients are seen by a doctor within the hour of arriving to the hospital, while others wait for 3–4 hours before they are seen. One patient describes her experience after waiting for 4 hours,

> There was a time I was about to leave, but then they called me in. I was literally getting up, and they called me up. [CAD-105-P]

One patient noted that he did not like going to the hospital because he spent so much of his childhood in that setting and would much rather stay in the comfort of his home. A common theme that arose from participants was the insensitivity to the pain experience and the lack of knowledge (N = 11, 38%) from nurses and doctors. Some participants felt they are not taken seriously, are overlooked, and need to explain their pain to the ER staff in order to get appropriate care. As previously mentioned, there is a preconception from staff in the hospital that black patients with sickle cell disease are only visiting the hospital to receive drugs. Additionally, a few patients (N = 4, 17%) specifically shared that they had to advocate for themselves in order for ER staff to understand their pain. One patient describes her experience with racial bias and how she changes her appearance in order to receive the proper care,

> Racial bias impacts how fast you get treatment … I get my crisis, at 4–5 [AM]. I'm in my sweatpants [and] sweatshirt. My hair's tied up … If you walk in looking like a bum, they're not going to take us serious. "Oh, she's here again for meds", this kind of thing … I realized … during my pain episodes, I'm dressing up and putting on makeup [and] fixing my hair, to be considered serious. And I realized other people are doing the same thing …That hinders treatment. [CAD-103-P]

Another patient shared how she preferred to visit the ER when someone else who can help advocate for her pain was present. This patient shares,

> If ever I'm in a crisis to the point where I cannot tolerate it … I usually don't want to go to the emergency room by myself. I'm not in a mental stage to advocate for myself. There is a component of having to explain yourself and explain sickle cell because … not all of your nurses are aware that there's a protocol in place. It's just a lot of energy … If I'm by myself, I would prefer going through the pain and then upping my dosage … in order to be able to stay through the pain at home [CAD-101-P]

These previous negative experiences have had a profound effect on the preference for care-seeking at home instead of a dedicated medical facility. Since these interviews took place during the COVID-19 pandemic, there was a fear for a lot of patients about getting COVID-19 or another infection (N = 6, 26%) if they visited the hospital, which contributed to the preference for care-seeking at home.

Subject Level Factors

We found the decision to seek care at a dedicated medical facility was slightly dependent on age, as more adolescent patients in a dyad reported care-seeking at a dedicated medical facility quicker than adult patients reported care-seeking. The decision of when to seek care was dependent on the caregiver's assessment of symptom severity and the caregiver's level of comfort caring for their child, as opposed to a specific elapsed time since symptoms began. One caregiver shared,

> He's taken in when I'm uncomfortable [When I am] like, "Okay, the pain's not subsiding. We've tried all these things". Then we go into the hospital, and it's more of a comfort level. It has nothing to do with anything else. I just don't have the machines to put monitors on him and verify. [CAD-212-C]

Notably, education level and disease severity did not relate to the decision to seek care at a dedicated medical facility.

A few patients noted they did not like going into the hospital because they do not have the time and they needed to continue with their everyday lives. Most of the adult patients (7/11, 64%) mentioned that they purposefully did not go to the ER because they needed to carry on with work or school, or they had a big exam or work-related task to do, and their teachers or employers would not be accommodating, even if they explained they were in an active pain crisis.

Most Important Patient/Caregiver Care-Seeking Factors

Given the factors mentioned around SCD symptoms and the treatment experience, participants were asked to rank the most important factors considered when treating a crisis, to further understand preference and drivers for care-seeking. Results from this ranking data can be seen in the Supplementary Data Table 4. Fifteen (65%) participants shared the most important consideration is the hospital experience, particularly with the treating medical staff and the care protocol implemented in the hospital. Given previous negative experiences, having a respectful, sensitive, and knowledgeable nurse or treating doctor at a dedicated medical facility was of particular importance for these patients when considering where to seek care for their pain crisis.

When analysing preferences within our sample, we found that most caregivers preferred to seek care at the hospital compared to their children who preferred to seek care at home due to the fear of missing school and the desire to be comfortable at home. While caregivers acknowledged the inconvenience for their child to be admitted to the hospital, most dyads (5/6, 83%) shared the decision to seek care outside the home ultimately relied on the caregiver's assessment of symptom severity. While only half of the caregivers (3/6, 50%) acknowledged the stressors of care-seeking at the hospital such as waiting time and stigma from hospital staff, almost all of the adult patients (9/11, 82%) acknowledged these same stressors, and in fact ranked these factors as important when considering where to seek care during a pain crisis. A few of these adult patients (5/11, 45%) shared they would rather seek care at home and only resort to the hospital when their symptoms are so severe that they have a fever, cannot walk, cannot talk, and/or are persistently crying.

Discussion

The study aimed to understand the SCD patient experience and the factors that contribute to care-seeking during a pain crisis. We specifically sought to identify patients and caregivers' attitudes and preferences when making decisions regarding care-seeking during a pain crisis.

Consistent with the literature, our study found acute pain crises in various parts of the body to be one of the primary manifestations of SCD, and poses a burden on social, emotional and physical domains.[11,12] One of the challenges when treating a pain crisis is that pain is a subjective, complex, and multidimensional experience, often presenting differently within patients and within pain crises.[13] Although patients have different thresholds and tolerances for pain, the symptom and pain severity was consistently noted among participants as a motivator for care-seeking at a dedicated medical

facility as opposed to care-seeking at home. This suggests that reliable and valid Patient Reported Outcome (PRO) measurements of pain would be important in research to understand care-seeking behaviours in the real world.

There are many factors that patients consider when deciding where to seek care, such as protocols in place, support systems in the hospital, and the administration of adequate pain medications. From our results, we found these attributes are not present across all hospitals and regions in Canada. In The Consensus Statement on the Care of Patients with Sickle Cell Disease in Canada published in 2018,[3] patients and caregivers are to be provided written instruction on how to deal with SCD pain and when to seek help. From our interviews, the majority of patients were not provided instruction on how to manage a crisis, and those that were provided some guidance, did not find the instruction helpful. Further, the Consensus Statement on the Care of Patients with Sickle Cell Disease in Canada[3] states that patients are to be assessed with priority upon arrival to outpatient management, a practice experienced by only few patients in our study. Some patients in our study shared that it takes them anywhere from one to four hours to be treated in the ER, which clearly falls outside of the recommendations from the CANHAEM consensus guidelines. According to one study on wait times for SCD patients in an ER in Ontario, adult patients waited an average of 90.6 minutes before being seen by a doctor while paediatric patients waited an average of 51.1 minutes.[14] Although there are apparent guidelines in place, the gaps are clear and from the patient perspective, our findings show that these gaps clearly drive care-seeking behaviours.

Most of the preferences for care-seeking at home were from adolescent and adult patients. In adolescent patients, their reasoning included emotional impacts from previous crises, such as feeling pressure from their teachers and peers due to missing school during a crisis. In particular, adolescents with SCD are expected to academically perform at the same level as their peers despite the physical and cognitive challenges that result from these crises. While adolescents shared a preference for care-seeking at home, the decision on where to seek care ultimately depended on the caregiver. It should be noted, however, that adolescent patients still appreciated the care they received at the hospital and were not as aware of the barriers to care, such as racial bias, as seen by their caregivers and adult patients. Adult patients shared a similar sentiment, in that their experience from previous crises led them to seek care in a dedicated medical facility only as a last resort.

This study adds to the literature on the misconceptions health providers have at dedicated medical facilities and the judgement on SCD patients being labelled drug-seeking when they arrive at a dedicated medical facility. Consistent with the literature, the majority of patients in our sample were of Black/African American descent. Similar to findings reported by Shapiro et al[15] and Waldrop and Mandry,[16] patients and caregivers in the present study reported experiencing stigma upon admission to the ER. While health providers have good intentions in keeping patients away from addictive painkillers such as opioids, this often leads to the mismanagement of patients and an unwillingness to provide the proper care. In addition, patients in our sample reported challenges in implementing a standard of care at their dedicated medical facility, sharing they needed to advocate for themselves and their child to be properly treated.

Our study is unique in that it gathered the care-seeking strategies and preferences of two age groups of patients living with SCD. Our study supports the literature on the challenges of adolescent to adult care, and the proper health-care transition that is required. SCD care transition refers to the entire period of preparation, transfer, and integration into adult care.[17] In our study, adolescent patients appreciated the care they received at their preferred medical facility (despite the impacts on their quality of life) whereas adult patients felt there were areas for improvement. This difference in perspective shared by both adolescent and adult patients represents a clear gap. While adult patients were not asked to share their transition from adolescent care, it is possible their preferences for care-seeking were influenced by insufficient preparation and integration from adolescent into adult care. Overall, it is clear from the present study that patients want to be understood and treated with respect when seeking-care, and that better education of hospital staff and consistent implementation of clinical guidelines would be useful to improve patient experience of care.

Benefits and Limitations

Identifying and recruiting patients and caregivers to participate in research for a rare disease such as SCD is challenging. Given such, the sample for this study is relatively small. This study did, however, benefit from the inclusion of dyads, to assess both caregiver and patient perspectives. While caregivers are able to provide a detailed description of their

management and care-seeking practices for their child, the child was able to speak to the impact on their day-to-day life. While caregiver and patient reports generally aligned, especially around symptoms and management strategies, we identified some differences in reported care-seeking barriers and factors, primarily due to the age of the adolescent patients. Our sample included patients and caregivers based only in Canada, and thus reflects limited geographic and cultural diversity. Consistent with the literature, our sample comprised a majority of Black/African American participants. Though no sex predilection for SCD exists, our sample consisted of predominately college-educated, female patients, which may not be representative of the whole SCD population.

Conclusions

The study results provide a rich description of the care-seeking preferences for pain crises in SCD patients in Canada, an area with a dearth of research in recent years. We found that many factors influenced care-seeking decisions, mainly the symptom and pain severity; institutional factors (waiting time, the implemented care plan, and the empathy or racial bias felt from medical facility); and subject-level factors (age and a flexibility in daily responsibilities). This highlights the importance of a unified care team that follows the same gold standard for pain crisis treatment, and that the learnings from patient experience of care be incorporated into SCD care protocols. Aside from the hospital care team, a supportive support system at the patients' school or workplace is especially important for patients to feel comfortable seeking care and missing school or work.

Further, the evaluation of adolescent to adult care transition is vital in order to meet the care-seeking needs of adult patients in particular. More research is needed to gather insights in a larger qualitative and quantitative study to understand trade-offs made in care-seeking for pain crises in SCD, so that all stakeholders in the care-relationship can be better informed and promote supportive relationships. As patients continue to consider care-seeking in the hospital, it could be valuable to understand how best patients would like to receive care in order to realise better health outcomes, provide relief from pain, and improve the overall quality of life in SCD patients.

Compliance with Ethics Guidelines and Informed Consent

This study was approved by Versiti (formerly Salus), of Austin, TX. Given the non-interventional nature of this qualitative study, no medical emergencies were anticipated as part of this research. Study objectives and methods of this study were explained to the participants during recruitment and verbal informed consent was obtained by all the participants at the start of the interview, before data collection. All verbal permissions confirming consent were audio-recorded from all participants and included in the transcription. All participants were informed that their personal information would remain anonymous in all written reports or publications. All participants were assured they could withdraw from the study at any time without impacting their treatment or medical care. No participants ended their participation or withdrew from the study. We confirm all procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1964, as revised in 2013.

Author Contributions

All named authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for this manuscript, take responsibility for the integrity of the work as a whole, and have given final approval to the version to be published.

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Disclosure

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d'Anémie Falciforme du Québec and aided in patient recruitment. The authors report no other conflicts of interest in this work.

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