ORIGINAL ARTICLE

A Needs Assessment of Persons With Sickle Cell Disease in a Major Medical Center in North Carolina

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BACKGROUND Sickle cell disease (SCD) is a complex disease associated with many complications and a shortened lifespan. In 2016, the National Heart, Lung and Blood Institute funded 8 centers in the United States to form the Sickle Cell Disease Implementation Consortium, with the goal of improving SCD care. The aim of our study was to describe SCD self-efficacy, pain interference, and barriers to care from the perspective of persons with SCD in the North Carolina center.

METHODS Persons with SCD, aged 15 and older were recruited from a large SCD center in North Carolina. Surveys, focus groups, and interviews were completed.

RESULTS Fifty-one people completed a survey, and 14 people completed an interview or focus group. Barriers identified in the survey included self-care barriers, misconceptions related to hydroxyurea (an oral medication that reduces rates of pain crisis), limited provider knowledge, and stigma. Concerning self-efficacy, participants reported that they were able to manage their pain symptoms most of the time. Pain interfered most with the ability to participate in social and day-to-day activities.

Common themes from the focus groups and interviews included misconceptions about hydroxyurea, pain, provider knowledge, stigma, co-management, transportation, and insurance. Recommendations to improve care included the use of case managers, utilization of treatment guidelines, individualized pain protocols, and effective co-management by providers.

LIMITATIONS Participants were recruited from 1 SCD center and may not be representative of the entire SCD population in North Carolina.

CONCLUSIONS Participants described many perceived barriers to care, and their responses suggest a need for improvements in patient hydroxyurea education, provider knowledge, and care coordination.

Sickle cell disease (SCD) is a genetic blood disorders predominantly affecting people of African ancestry [1, 2]. In the United States, 1 out of every 356 African Americans is born with SCD, and 1 out of 13 African Americans carries the SCD trait [1, 2]. Approximately 5,500 individuals with SCD live in North Carolina [3]. The disease is associated with many complications including pain [4]. Interventions such as newborn screening, vaccination, and hydroxyurea (an oral medication that reduces rates of pain crisis) use have led to an increased life expectancy over the past twenty years [4]. In North Carolina, the scale up of these interventions, particularly newborn screening, was facilitated by the NC Sickle Cell Syndrome Program [5]. The program consists of 6 comprehensive medical centers, community-based organizations, regional counselors, and state staff [5]. Despite these interventions, individuals with SCD still encounter several barriers to care [6–8]. Not all individuals with SCD have access to quality care, particularly at the primary care level, and as a result, may experience poor quality of life and a shorter life span [9].

The American Society of Hematology (ASH) and the National Heart Lung and Blood Institute (NHLBI) provided national guidelines and recommendations for the management of acute and chronic SCD complications, as well as care coordination among providers [10, 11]. However, those recommendations, particularly those for care coordination, are difficult to implement. In order to improve the implementation of the recommendations and address barriers to SCD care, the NHLBI funded the 8 centers to participate in the Sickle Cell Disease Implementation Consortium (SCDIC) [12]. The 8 centers represent academic medical institutions and multi-disciplinary, community-based implementation science research teams [12]. The consortium conducted a needs assessment of individuals with SCD. This needs assessment will inform the development and testing of interventions to improve the outcomes for persons with SCD. This paper reports results from the needs assessment conducted in North Carolina and describes SCD self-efficacy, pain interference, and barriers to care from the perspective of individuals with SCD.

Methods

Study design and setting

A descriptive, multi-methods study design using surveys

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and a combination of focus groups and in-depth interviews were used to conduct the needs assessment. The study took place in a large comprehensive sickle cell center in Durham, North Carolina. The study site serves over 700 adults with SCD. Almost all (approximately 96%) of the individuals who receive care at the comprehensive sickle cell center have medical insurance.

Participant recruitment

Participants were eligible for enrollment if they were aged 15 years old or older and had a self-reported diagnosis of SCD. Participants were recruited through flyers that were distributed to SCD support groups in North Carolina or were introduced to the study by their health care providers during their routine clinic visits. In-person recruitment at support groups and clinics was done by study coordinators. Participants were approached and given the option to complete the needs assessment survey and then opt in or out of being interviewed at a later date.

Ethical approval was obtained from Duke University Institutional Review Board prior to data collection. Written informed consent was obtained from eligible and willing participants. We received parental consent and assent for participants aged 15-17 years. Personal identifying information was not collected in order to preserve participant confidentiality.

Measures

Surveys

The survey was developed by the SCDIC needs assessment subcommittee composed of members from all 8 centers. Members included hematologists, advanced practice providers, and researchers with expertise in SCD. The survey consisted of a combination of validated instruments that assessed demographics and SCD genotype, perceived sickle cell self-efficacy, pain interference, and barriers to SCD care. SCD self-efficacy was measured using the SCD self-efficacy scale [13]. This 9-item scale includes questions relating to participant perceptions of their ability to function on a day-to-day basis and to manage their SCD-related symptoms [13]. The scale was developed using a community-based sample of adults with SCD and found to be reliable (Cronbach alpha 0.89) and valid [14]. Pain interference was measured using 4 items from the Patient-Reported Outcomes Measurement Information System (PROMIS), a set of 29 person-centered measures to evaluate and monitor physical, social, and mental health in adults and children [14]. PROMIS item banks were developed using comprehensive literature reviews, focus groups, cognitive interviews, psychometric testing, and validity testing [15]. Clinical validation of the pain interference questionnaire has been conducted in diverse clinical populations such as cancer, low back pain, and rheumatoid arthritis [16, 17]. SCD specific barriers to care were assessed using 8 multiple-choice questions, a checklist, and 1 comment area where participants could respond using additional text. The checklist has demonstrated face validity and test-retest reliability (Pearson r = 0.74, $P < .05$) [14]. Participants could select more than 1 of the barriers provided in the following categories related to SCD: provider knowledge and attitudes; hydroxyurea use; social and family caregiver support; access and accommodation within places you get health care; barriers for individuals; insurance; transportation; and access to service.

Focus groups and in-depth interviews

A semi-structured interview guide was developed and agreed up by members from each of the consortium sites. The following main areas of care were determined to be the focus of the interviews: access to primary and specialty care, hydroxyurea use, and beliefs and practices related to pain control. The interview questions were developed by members of the SCDIC Needs Assessment Committee based upon expert knowledge and current literature. Guiding questions included: 1) What kind of health care providers do you see? (or who do you see for your health care?); 2) Tell us about hydroxyurea, whether you use it, decided not to, or just what you have heard about it; and 3) Tell me about the pain you experience and how you manage it.

Procedures

The surveys were administered in person by the study coordinators, were completed electronically, and took approximately 45 minutes to complete. The majority were completed in the clinic immediately after consent was obtained.

Ten in-depth interviews and 2 focus groups with 2 participants each were conducted. Each interview lasted approximately 60 minutes. The planned study design was only focus groups, but due to scheduling conflicts caused by acute medical complications, hospitalizations, and unexpected travel plans, the study design was amended to also include individual telephone interviews.

Data analysis

Survey data were entered electronically using REDCap data capture software [18]. SAS (version 9.4) statistical software was used to obtain descriptive statistics expressed as frequencies, means, and standard deviations.

The interviews were recorded and transcribed verbatim. Four members of the research team validated the accuracy of the transcription. All transcripts were coded and analyzed using QSR NVivo (version 11). A codebook was developed based on a priori codes adapted from literature on SCD and emerging themes from the transcripts. Three members of the research team (N.C., E.B., R.M.) independently coded 10% of the interview transcripts. Coding disagreements were resolved through repeated discussions until consensus was achieved. A weighted kappa value of 0.95 was achieved. The remainder of the transcripts were coded using the final agreed upon coding structure.

Results

Sixty-five people were screened, 7 people were lost to contact, and 58 consented to participate in the study. Two focus groups (N = 4) and 10 individual interviews were conducted, and 51 surveys were completed. Participant demographics are highlighted in Table 1. Majority of the participants were non-Hispanic, African American, and had SS genotype.

Survey results

Self-efficacy. Levels of self-efficacy are reported in Table 2. Most participants reported they were sure that they could cut down on their pain, could manage day-to-day activities, and could do something to help themselves when they were feeling sad. Most also reported they could manage symptoms to enjoy life and deal with the frustrations of having SCD. Participants were less sure about the ability to reduce SCD pain using non-pharmacologic methods. However, they were least sure about their ability to control how often or when they get tired or how to reduce SCD pain from interfering with sleep.

TABLE 1.
Participant Demographic Characteristics (Surveys, Interviews and Focus groups)

Variable	Survey N = 51	Interiews/ Focus Groups N = 14
Age		
Mean (SD)	30.7 (10.6)	32.5 (5.7)
Median	28	32
Minimum-Maximum	16-66	25-43
Missing, n	6	1
Ethnicity/Race, n (%)		
Non-Hispanic/Black	45 (88.0)	13 (93.0)
Hispanic/Black	3 (6.0)	1 (7.0)
Don't know/Black	1 (2.0)	
Missing, n	2 (4.0)	
Gender, n (%)		
Male	26 (52.0)	5 (36.0)
Female	24 (48.0)	9 (64.0)
Missing, n	1 (2.0)	
Insurance, n (%)		
Private insurance only	9 (18.4)	2 (14.3)
Medicare only	4 (8.2)	2 (14.3)
Medicaid only	15 (30.6)	4 (28.6)
Private Insurance + Medicaid	2 (4.1)	0
Private Insurance + Medicare	1 (2.0)	1 (7.1)
Medicare+ Medicaid	14 (28.6)	3 (21.4)
Other government program	1 (2.0)	0
State sponsored insurance	1 (2.0)	0
Medicare + Other government program	1 (2.0)	0
Don't know	1 (2.0)	0
Missing, n	2 (4.0)	2 (14.3)
Genotype, n (%)		
SS	38 (76.0)	
SC	8 (16.0)	
S Beta 0 Thalassemia	1 (2.0)	
SB+	2 (4.0)	
SS Alpha Thalassemia	1 (2.0)	
Missing, n	1	

Note. Missing = participants who provided no response to the survey item.

Pain interference. The degree of pain interference, using PROMIS, is reported in Figure 1. Pain interfered most with the ability to participate in social activities, day-to-day activities, and work around the house. However, participants responded that pain interfered "quite a bit" or "not at all" with household chores.

Survey reported barriers to care. Barriers to care identified in the surveys are provided in Table 3. Pain was the most frequently selected barrier, followed by being tired. Forgetting to take medication was the main barrier to hydroxyurea use. Provider-related barriers were a lack of knowledge and experience with SCD, not being seen quickly enough when in pain, and lack of belief that they were in pain. Transportation was frequently endorsed as another barrier to care; poor access to public transportation and not owning a vehicle were the most common transportation barriers. Social-related barriers included dependence on family assistance in daily chores and activities and caregiver burnout. System-level barriers included difficulty communicating with staff and finding the health care system hard to navigate.

Themes from interviews and focus groups

Major themes that emerged throughout the interviews were pain, provider knowledge, stigma, hydroxyurea, co-management among health care providers, transportation, and insurance. Participants recruited in the clinic and support groups has similar perspectives on the major themes.

Pain. Participants had varied descriptions of pain: "pulsating," "throbbing pain," "a burning sensation where bones felt like they were on fire," "being stabbed from my face down," and "everything hurts and nagging." Pain crises were triggered by several things, such as cold weather, stress, or normal daily activities, and sometimes had no trigger at all. Pain was compounded by other co-morbidities, such as avascular necrosis and arthritis, and required multiple strategies to alleviate. Participants expressed their desire to live a pain-free life, were willing to do anything non-invasive, and only went to the hospital when pain-mitigating strategies failed. Pain affected their quality of life and personal and professional relationships.

Provider knowledge. Many participants emphasized the importance of provider SCD knowledge. Several participants reported provider lack of knowledge as a barrier to having a primary care provider (PCP) or having a good relationship with their PCP.

> *"I don't have a primary care physician. I've been looking for one but I think the biggest barrier for that is trying to vet the primary care doctor you know well enough to see if they know enough about sickle cell."*

TABLE 2.
Perceived Sickle Cell Self-Efficacy

Sickle Cell Self Efficacy	Frequency (%)	Sickle Cell Self Efficacy	Frequency (%)
How sure are you that you can do something to cut down on most of the pain you have when you have a pain episode?			
Not at all	2 (4)	Not Sure	8 (16)
Neither	8 (16)	Sure	23 (46)
Very Sure	9 (18)	Missing, n	1
How sure are you that you can keep doing most of the things you do day-to-day?			
Not at all	0 (0)	Not Sure	14 (28)
Neither	5 (10)	Sure	23 (46)
Very Sure	8 (16)	Missing, n	1
How sure are you that you can keep SCD pain from interfering with your sleep?			
Not at all	12 (24)	Not Sure	16 (32)
Neither	6 (12)	Sure	13 (26)
Very Sure	3 (9)	Missing, n	1
How sure are you that you can reduce your SCD pain by using methods other than taking extra medication?			
Not at all	5 (10)	Not Sure	19 (38)
Neither	7 (14)	Sure	13 (26)
Very Sure	6 (12)	Missing, n	1
How sure are you that you can control how often or when you get tired?			
Not at all	11 (22)	Not Sure	16 (33)
Neither	7 (14)	Sure	13 (27)
Very Sure	2 (4)	Missing, n	1
How sure are you that you can do something to help yourself feel better if you are feeling sad or blue?			
Not at all	1 (2)	Not Sure	5 (10)
Neither	5 (10)	Sure	28 (56)
Very Sure	11 (22)	Missing, n	1
Compared to other people with SCD, how sure are you that you can manage your life from day-to-day?			
Not at all	0 (0)	Not Sure	2 (4)
Neither	7 (14)	Sure	22 (44)
Very Sure	19 (38)	Missing, n	1
How sure are you that you can manage your SCD symptoms so that you can do the things you enjoy?			
Not at all	1 (2)	Not Sure	7 (14)
Neither	6 (12)	Sure	21 (42)
Very Sure	15 (30)	Missing, n	1
How sure are you that you can deal with the frustration of having SCD?			
Not at all	4 (8)	Not Sure	9 (18)
Neither	7 (14)	Sure	17 (35)
Very Sure	12 (25)	Missing, n	2

Note. Missing = participants who provided no response to the survey item.

This was also relevant for those seeking care from different specialists. Participants felt they were limited in the providers they could trust with their care due to unfamiliarity with SCD. Conversely, participants felt experienced providers facilitated care.

> *"He had prior knowledge of dealing with people with sickle cell disease and [...] I believe he said he's attended seminars and he's met doctors from the organization that have helped him in his medical career as far as dealing with sickle cell patients."*

Stigma. Many participants reported that stigma was a barrier to care and describe experiences that illustrated the wide-ranging effect stigma had. Stigma of SCD removed their individuality by categorizing them as drug seekers. One participant described the desire to stop their pain, rather than a desire to "get high" as the motivator for the behaviors that have been characterized as "drug seeking."

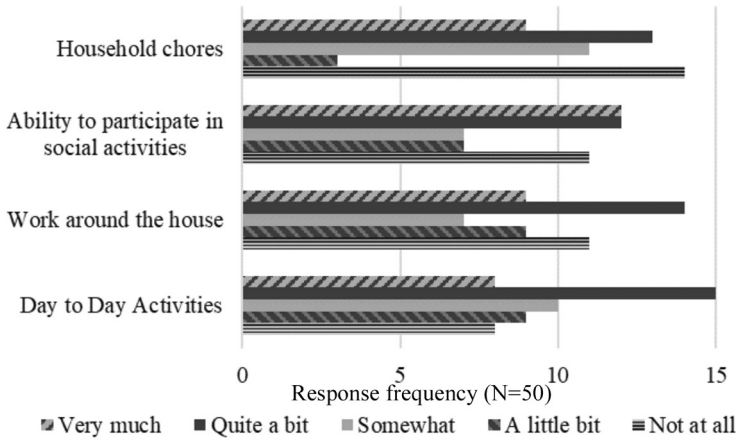
> *"You are drug seeking but not because you want to get high. Not because you want to feel good. [...] No, you want the pain to stop."*

The stigma of having SCD and being categorized as drug seekers were described by several participants as significant life experiences. One participant described how the drug seeking stigma led to an encounter with the police when they were filling a prescription at the pharmacy.

> *"I was sitting there waiting and these two officers came up to me and asked me was I [redacted] and I had to show identification and they said that they got a call that I was trying to fill a barred prescription and so I had to end up like calling my doctor and having the pharmacy call [redacted]."*

Hydroxyurea. Participants described a variety of ways in which they manage their disease, including hydroxyurea. Several participants described their parents as the primary

FIGURE 1.
Level of Pain Interference in Day-to-day Activities, Household Chores, and Social Activities



Note. X-axis represents the activities affected by pain interference; y-axis represents the participant response frequency.

decision-maker in managing their hydroxyurea during childhood.

> *"I basically just did what my mom told me to do which was take these pills. And just take your pills and make sure you take your pills, so. It wasn't until I got older. I mean my mom after we reached a certain age let us be responsible for ourselves. Like my brother chose not to take it so she said that's your body, that's you."*

Participants reported various misconceptions about hydroxyurea, including that it is a chemotherapeutic agent that was still undergoing drug trials and expressed concern about being a "guinea pig."

> *"One time I was in the hospital and this nurse was handling it with gloves and I was like why you got gloves on to touch this medicine? She was like, oh because it's a chemo drug and I'm like, I'm on chemo?"*

Participants described several aspects of treatment with hydroxyurea that were difficult for them to manage and sometimes led to their stopping or refusing to start the medication. Challenges included determining therapeutic dosing, drug side effects, daily administration, and the need to stay on hydroxyurea for a lifetime. Only a couple of participants provided recommendations related to hydroxyurea adherence. One patient described rarely missing a dose and attributed this to having a very involved health care provider, who at every visit asks what doses he is taking, how often, and if doses were missed.

Care coordination. Participants reported that lack of communication between providers was often a barrier to care. This was particularly relevant for primary care providers (PCPs). Those that reported that they did not have a primary care provider acknowledged the importance of having a PCP, and some expressed guilt for lacking a PCP. Some participants reported the reason they did not have a PCP was due to previous negative experiences, stating that their former PCP did not work well with their hematologist, leading them to become distrusting of new providers.

Conversely, the presence of care coordination between health care providers was a facilitator to care. Participants reported that communication and coordination between PCPs or specialists and their sickle cell team was an advantage. One participant reported that although they do not see their PCP often, the relationship they had was positive, attributing this to the primary care provider's willingness to communicate.

Transportation and insurance. Many reported that transportation was currently, or previously, a barrier to attending appointments, particularly when they lived longer distances from their clinic. Participants often relied on their family members or public transportation to get to appointments. One participant described their inability to attend appointments due to lack of transportation as troubling since they felt embarrassed about the impression this left with providers.

> *"Relying on people for rides is a headache or if you don't have money to get on the bus it's a headache so it's like when you have doctor's appointments and you can't get there it's like you got a bad face because you didn't come to your appointments."*

While all participants reported that they had insurance, several had experienced lapses in insurance at one point in time or were unable to get coverage for certain procedures. Several participants described concerns related to cost from co-payments and identified this as a barrier to obtaining care.

Recommendations. Participants often spontaneously provided recommendations during the interviews on how to improve SCD care. Subthemes for recommendations were identified and are summarized in Table 4. They included provider knowledge and empathy; care coordination and follow-up; individualized pain protocol; complementary resources; transport and treatment costs; and addressing misconceptions about hydroxyurea.

Discussion

The purpose of this study was to describe SCD self-efficacy, pain interference, and barriers to care from the perspective of individuals with SCD in North Carolina. Our study revealed an interplay of varied SCD care needs that can be broadly classified at the individual, social, provider, and system level. We identified opportunities for health care providers to address some of the individual needs.

Individual level factors

Individual level factors that influence SCD care included pain, transportation, and perception of hydroxyurea. Pain was the most dominant barrier to care identified in the surveys and interviews. Details on how pain affects participants varied. For example, over half of survey participants indicated that they were "not at all" sure or "not sure" they could control their SCD pain so that it would not interfere with sleep. Pain-associated sleep interference in adults with SCD has been reported as comparable in severity to other chronic illnesses and as a high priority concern in prior studies [19–24]. Health care providers have an opportunity to explore how pain impacts sleep for all individuals with SCD.

Lack of transportation to the health facilities, particularly to the regional or university-based hospitals, led to frequent missed appointments with subsequent exclusion of participants by the clinics. Evidence-based solutions targeted towards reducing the distance between the patient and health care provision include the use of community health workers, satellite clinics or mobile health centers, and telemedicine [25–27]. Community health workers have been proven to have a positive effect on health outcomes [27]. Further studies are required to determine if satellite clinics and mobile health centers provide the same level of comprehensive care as the regional and university-based hospitals. The more recent expanded use of telehealth and telephone visits provides an additional opportunity to improve care.

Poor adherence and utilization of hydroxyurea in SCD had been identified in literature [28,29]. Participants indicated that forgetting to take medication was the biggest barrier to hydroxyurea adherence [30]. Our qualitative data indicated a high degree of skepticism related to the effectiveness of hydroxyurea and persistent misconceptions about hydroxyurea being an experimental chemotherapeutic drug. Our qualitative data also suggest that hydroxyurea use in childhood is primarily determined by parents and that in childhood, participants were unlikely to question why they took hydroxyurea. However, as adults, participants indicated that they had chosen not to take hydroxyurea because of concerns related to side effects, difficulty in the dosing regimen, and not wanting to take a daily medication. These results indicate the complexity of reasons why hydroxyurea adherence is typically low. Providers should explore an individual patient's

TABLE 3.
Barriers to Sickle Cell Disease Care Checklist

Barriers Category	Frequency
Barriers related to SCD	
I am in pain	28
I am tired	26
Frustration or anger	22
Worry or fear	20
No barriers	11
Lack of confidence	10
It is embarrassing	7
Concerned about the costs	6
Hard to be assertive	6
Provider knowledge and attitudes	
No barriers	23
Not enough experiences with or knowledge about SCD	19
Not seen quickly enough when in pain	16
Don't believe I am in genuine pain	13
Accused of drug seeking	12
Treated differently from other patients	10
Communication between me and provider is difficult	9
No appreciation of how knowledgeable I am about SCD	3
Hydroxyurea use	
No barrier	22
I forget to take the medication	10
Other barrier	8
Don't like thinking about SCD when I am well	6
Worried about the side effects	6
Don't know enough about the medicine	3
It is hard to take the medicine at the right time	3
I tried it and it did not work	3
Heard some scary things about this medicine	2
I don't like the frequent blood tests	2
I don't like the frequent clinic visits	2
Not interested in taking another medicine	2
Doctor does not recommend it	2
There are other things going on in my life that are more important than taking this medicine	1
Social, family, and caregiver support	
No barriers	33
I need help with daily chores/activities	7
The people who take care of me or give me support are burned out	7
I am socially isolated	5
There are other things in my family that are more important than my health care	4
I am burned from providing care to others	4
It is hard to make appointments because it is hard for me to find childcare during	2
I do not have enough support	2
Access & accommodation within places you get health care	
No barriers	31
Wait in health care office is too long	11
Health care providers' hours are not convenient for me	7
Places for me to go learn how to stay well are not close by or easy to get to	7
I could not get an appointment	2
The paperwork I have to fill out is too much	2

TABLE 3. CONTINUED
Barriers to Sickle Cell Disease Care Checklist

Barriers Category	Frequency
Barriers for individuals	
No barriers	35
Staff is hard to talk to	5
I don't understand the system or find it too hard to work through	5
Medical system is very confusing to me	3
Staff is hard to understand	3
I missed appointments because of my memory problems	3
Other health problems have higher priority	2
It is hard to follow up on care (e.g., by going to the pharmacy, taking medicines at the right time or making follow up appointments)	2
Don't know what to do to stay healthy	2
I am not interested in getting SCD care	1
I don't know enough about the SCD care that I need	1
Hard for staff to get a hold of me (e.g., I move a lot or don't have a phone)	0
Medical instructions are hard to follow	0
Insurance	
No barriers	38
My insurance will not cover needed services if I have to go to a different county	3
Getting reimbursement for some services is hard	3
Does not cover medicines or my copay are too high	3
Does not cover services that will keep me well	3
Doesn't cover services I need	3
My insurance paperwork too hard to fill out	2
It takes too long to get approval for care	2
My insurance doesn't allow me to go where I want for service	2
No insurance	0
Transportation	
No barriers	34
I do not have a vehicle	8
Public transit is not easy to get to	6
Transportation costs too much for me	3
I can't get transportation	2
I do not have access to a vehicle	1
Access to service	
No barriers	45
I can't get care because the health care provider's office is too far away	2
I don't know where to get care	2

Note. Missing = participants who provided no response to the survey item.

barriers to taking hydroxyurea and target patient education accordingly. There is also an opportunity to emphasize education of children with SCD about hydroxyurea and continuing this education throughout adulthood [31].

Social level factors

Social level factors included stigma and family support. Stigma has been identified in several studies of barriers to SCD care [32,33]. Despite the recognition that stigma is a common barrier to care, there are few interventions that have been developed to reduce stigma in this population, and these have focused on knowledge/education of providers about SCD and on improving provider communication techniques.

Family involvement in care was identified during interviews as key in assisting participants meet their health needs. Participants identified caregiver burn out and needing help with daily chores/activities as barriers. Participants were not directly asked about caregiver needs or concerns during the interviews; however, this may be an important area to explore in future qualitative studies.

Provider level factors

Provider level factors included knowledge and co-management strategies. Participants frequently perceived a lack of knowledge about common SCD complications by their providers as a barrier to care. Despite the availability of national treatment guidelines from the ASH and NHLBI, few PCPs are aware of or follow those guidelines [34,35]. Health care providers could therefore benefit from targeted training on how to manage pain using the ASH and NHLBI guidelines. Additionally, methods to make these guidelines readily accessible to a wide variety of providers could improve management of SCD.

In addition to provider knowledge, the long-term management of individuals with SCD requires frequent follow-up care with SCD specialists such as hematologists, as well as a care model in which patients are co-managed with PCPs [36, 37]. Coexisting chronic diseases, such as diabetes and asthma, are often underevaluated by hematologists who are not primarily required to manage these conditions. A co-management care model is therefore needed to foster partnerships between SCD specialists (hematologists) and PCPs in order to manage the individual with SCD in a holistic manner [36]. In addition, comanagement presents a good opportunity to help address transportation barriers by allowing patients to receive general health care with their PCP and more specialized care from their hematologist.

System level factors

Participants reported frequent difficulties navigating a complex health care system and insurance-related barriers. Participants described difficulty coordinating numerous specialist appointments needed for their disease management and long-waiting times in scheduling those appointments. These barriers could be addressed by increasing the use of telephone and telehealth visits as well as care managers.

Similar to other studies, participants were mostly Medicaid insured and reported high costs of insurance copayments [38–40]. Concerns about lapses in insurance and lack of continuity in care due to changes in coverage were common. Incorporating care managers into provider practices could be one method of helping patients address insurance concerns. Every patient with Medicaid is assigned a care manager in North Carolina [41].

TABLE 4.
Recommendations for Improved Care

Category	Recommendations
Provider knowledge and empathy	1. Improvement in sickle cell disease knowledge and management of painful crisis.
	2. Implementation of guidelines that ensure standardized care in the hospital.
	3. Empathy and compassion from providers.
	4. Stigma mitigating strategies that will ensure less bias.
Care coordination and follow-up	1. Improvement in communication among clinicians or specialists.
	2. Establishment of follow up appointments with relevant specialists.
	3. Facilitated provision of prescriptions, particularly for opioids.
Individualized pain protocol	1. Easy access to electronic medical records by the patient and provider.
	2. Individualized pain protocols listing drug types and dosages that alleviate pain.
Complementary resources	1. Non-medical personnel such as case-managers and social workers, who will facilitate adherence to clinic appointments.
	2. Psychosocial support by chaplains and patient champions or advocates with a similar disease burden.
Transport and treatment costs	1. Reduction in treatment costs for uninsured patients.
	2. Provision of transport to medical appointments.
Address misconceptions about hydroxyurea	1. Classes in the sickle cell clinic.
	2. Online tutorials.
	3. Success stories on the effectiveness of hydroxyurea.

Limitations

There were limitations to our study. All participants were recruited from 1 center and had medical insurance, thus they may not be representative of the entire SCD population in North Carolina or the United States. However, despite being from a large SCD center and having medical insurance, participants still expressed many barriers to receiving care. It is possible that these barriers are even more problematic for individuals who are uninsured or not able to access care in a comprehensive SCD center. Despite these limitations, this study does possess several strengths. We utilized multiple methods, inclusive of surveys and in-depth interviews, that provided an in-depth analysis of the factors influencing quality care provision [42]. Findings from this paper will inform interventional studies targeting individuals with SCD who are affiliated and not affiliated to health care throughout North Carolina.

Conclusion

Participants described many perceived barriers to care and identified a need for interventions to address hydroxyurea adherence and stigma, methods to improve provider knowledge in addressing the complex health care needs of SCD, and a co-management model that facilitates better delivery of care. Greater attention to the effects of SCD on the family (e.g., caregiver burnout) and identification of the resources required to support those families are also needed. NCMJ

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