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"I can't cry on cue": Exploring distress experiences of persons with sickle cell

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ABSTRACT

Persons with sickle cell disease exhibit high levels of distress, which has been documented with validated measures. However, there has been little qualitative investigation of the sources of distress in the lives of persons with sickle cell or the strategies they use to manage different sources of distress. Our study sought to: (1) identify sources of distress for persons with sickle cell, (2) explore management strategies for different sources of distress, (3) inform content development of the future mobile phone application, and (4) incorporate patient voices in the design and development phases of the future mobile phone application. In this manuscript, we present findings for the first objective. Using convenience sampling, we recruited participants with sickle cell (n = 11) from a home visit program at The Ohio State University Wexner Medical Center between February and July 2021. One team member conducted one-on-one semi-structured interviews with participants. We coded and analyzed all transcripts. Participants identified clinical encounters in the emergency department and intermediate care center to manage acute pain flares as the most profound source of distress. Our analysis identified four themes: (1) Pain has performative features, (2) Stigma and racism surround care, (3) Sickle cell is a neglected disease, and (4) Participants lack control over their pain management plan. Researchers may wish to consider how these settings can foster distress, and providers may wish to adopt participant recommendations to reduce distress associated with seeking pain treatment in acute care settings.

1. Introduction

Sickle cell disease is associated with chronic pain that can be difficult to treat. It is also characterized by high levels of emotional or psychosocial distress (Edwards et al., 2009, 2014; Treadwell et al., 2015, 2022). A robust body of literature has investigated sickle cell pain, healthcare utilization, distress associated with sickle cell, and the experiences of persons with sickle cell in managing their condition.

Most persons with sickle cell experience painful crises, but the frequency of these crises varies considerably. Moreover, not all persons with sickle cell suffer from chronic pain or use opioid medications daily (Karafin et al., 2019; Prince et al., 2023). However, chronic pain has a significant impact on quality of life (Matthie et al., 2020; Santos et al., 2021), high opioid use, and opioid tolerance. These are of great concern to treating physicians and policymakers. The current evidence indicates the need to improve our understanding of the subjective experience of pain in the subset of persons who have chronic pain. This can shed light on the complex drivers of healthcare utilization and opioid use in persons with sickle cell who also have chronic pain, and help identify ways to improve their care.

The Pain in Sickle Cell Epidemiology Study (PiSCES) studied many aspects of acute and chronic sickle cell pain prospectively for two years (July 2002–August 2004), reporting on correlations between pain and health service utilization, site of primary health care, quality of life, individual coping strategies, depression, anxiety, and substance abuse among other outcomes (Aisiku et al., 2007; Citero Vde et al., 2007;

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Levenson et al., 2007; McClish et al., 2005, 2009, 2017; Smith et al., 2005, 2008, 2018; Sogutlu et al., 2011). Central sensitization (nociceptive hyperexcitability that heightens pain) is associated with higher self-reported clinical pain, along with vaso-occlusive crises and negative mood (Campbell et al., 2016). Chronic social defeat, such as repeated failures to obtain adequate pain relief, can enhance central sensitization, lowering the threshold for pain perception and amplifying negative emotions associated with pain (Biltz et al., 2022).

While providers are also concerned about the rate of healthcare utilization in this population, persons with sickle cell average just one hospital admission per year (Carroll et al., 2009). An investigation of data from five years of inpatient hospital claims for a managed care organization in Baltimore, MD found that only a subset of this population averaged more than four hospital admissions per year and this subset was the same age as low utilizers. Only a small proportion of this subset maintained high utilization patterns over the five-year claims period. A subsequent study reinforced the finding that persons with sickle cell who have high utilization of healthcare services moderate quickly (Carroll et al., 2011). Moreover, persistent high utilization by persons with sickle cell is associated with new substance use and mood disorder diagnoses rather than complications from the condition (Carroll et al., 2011). A multicenter, prospective cohort study describing acute care utilization in adults, found multiple predictors of higher utilization (Lanzkron et al., 2018). These include unemployment, having chronic pain, undergoing chronic transfusion therapy, a history of stroke, and receiving disability or Medicaid benefits.

Distress and pain are closely associated and can be co-occurring, but their relationship is complex. There is evidence that emotional reactions to pain are predictors of psychological distress (Edwards et al., 2009). Several studies have shown a positive association between (1) perceived discrimination or mistreatment, and (2) pain burden in patients with sickle cell disease (Ezenwa et al., 2017; Haywood et al., 2014). Perceived stress is associated with pain intensity in adults with sickle cell (Porter et al., 1998), and anxiety, rather than depression, and has been identified as the mediator of the relationship between stress and pain (Robbins et al., 2019).

Distress in persons with sickle cell has been measured using PROMIS (Patient-Reported Outcomes Measurement Information System) and ASCQ-Me (Adult Sickle Cell Quality-of-Life Measurement System). The PROMIS system assesses aspects of well-being including pain, physical and mental health, and social functioning (Dampier et al., 2016a, 2016b; Hays et al., 2018; Pope et al., 2021; Reeve et al., 2007). ASCQ-Me captures cognitive, emotional, physical, and social impacts, and pain episodes for persons with sickle cell (Keller et al., 2014, 2017; Treadwell et al., 2014). Distress in persons with sickle cell is associated with lower mental and physical quality of life, disrupted sleep, and high levels of discrimination, suicidal ideation, anxiety, depression, and extreme pain (Abdallah et al., 2020; Edwards et al., 2009; Knisely et al., 2021; Treadwell et al., 2011, 2015; Wallen et al., 2014).

Extensive qualitative and mixed-methods literature from the last two decades offers insight into the pain experiences of persons with sickle cell, and the challenges of managing this condition. The complexity of sickle cell pain was the focus of the earliest qualitative investigations, offering insight into how the experience of pain for persons with sickle cell differs from other types of pain, along with the limitations of pain scales in conveying pain quality and intensity (Adegbola et al., 2012; Booker et al., 2006; Coleman et al., 2016; Jacob, 2001). The necessity of self-management is another theme in this literature. Research highlights the challenges and burdens that self-management poses (particularly for younger patients), the use of multiple methods of pain self-management, and the time-consuming work of self-management for persons with sickle cell, who must organize their lives around self-care (Elahi et al., 2017; Matthie & Jenerette, 2017; Matthie et al., 2015, 2019; Thomas & Taylor, 2002). Experiences of stigma are well-documented, with extensive evidence that persistent racial bias has fostered mistrust of providers and the healthcare system (Haywood et al., 2010; Power-Hays & McGann, 2020; Tsyvkin et al., 2015; Young et al., 2020; Zaidi, 2021). Persons with sickle cell report experiencing stigma in clinical encounters related to being persons of color and having pain, with providers labeling them as drug-seeking (Bulgin et al., 2018; Liederman et al., 2021; Renedo et al., 2019; Sinha et al., 2019). Past negative experiences of provider treatment in the emergency department may discourage or delay subsequent attempts to seek emergency treatment (Abdallah et al., 2020; Abu Al Hamayel et al., 2021; Crego et al., 2021; Elahi et al., 2017; Jenerette et al., 2014; Thomas & Taylor, 2002; Tsyvkin et al., 2015; Young et al., 2020).

Despite this extensive literature, there has been little qualitative investigation of how persons with sickle cell experience and manage distress in their daily lives. We undertook a qualitative study with four objectives: (1) identify sources of distress for persons with sickle cell; (2) explore their strategies for managing different sources of distress; (3) inform content development of a future mobile phone application with these findings; and (4) embody patient voices in the design and development phases of a future mobile phone application.

The purpose of the future mobile phone application in development is to deliver a mindfulness-based pain management program tailored to persons with sickle cell. There is growing evidence for the acceptability and feasibility of mobile health interventions for adults with sickle cell, and evidence of their modest impact for pain and symptom management (Badawy et al., 2018; Jonassaint et al., 2015; Shah et al., 2014). Mindfulness is often defined as moment-to-moment awareness (Creswell, 2017; Kabat-Zinn, 2005). Mindfulness-based interventions and approaches can impact quality of life, anxiety, depression, chronic pain, and coping patterns (Brintz et al., 2020; Gilbertson & Klatt, 2017; Grossman et al., 2004; Kabat-Zinn et al., 1985; Schuman-Olivier et al., 2020). A single study has shown acceptability and feasibility of a mindfulness-based intervention for persons with sickle cell (Simmons et al., 2019). However, to our knowledge, no mobile phone applications deliver mindfulness-based pain management programs that help persons with sickle cell understand the sources of distress in daily life and their impact on pain.

To inform the content and design of a future mobile phone application, we conducted a qualitative study with persons with sickle cell to understand their experiences of distress and their self-management strategies. Our goals were also to understand their aptitude for mobile applications and social media, their design preferences for mobile applications, and their needs for a mobile pain management tool. In this publication, we present findings from our analysis of interview data for distress experiences and management strategies only. We closely examine one source of distress that participants identified as the most powerful. Development of the content and design of the future mobile application will be determined by our interview analysis, and prototype testing, currently underway. We will discuss content and design of the future mobile application in another manuscript.

2. Materials and methods

2.1. Study design and recruitment

We developed a meaning-centered qualitative design with one-on-one, semi-structured interviews for this study. The team developed an interview guide with nineteen questions in seven domains: (1) definitions of distress; (2) distress experiences; (3) role of identity in distress; (4) distress self-management strategies; (5) attitudes towards complementary therapies and mindfulness; (6) mobile application use and preferences; and (7) other information.

Using convenience sampling, the team recruited persons with sickle cell disease (n = 11; mean age = 43 years) from a home visit program at The Ohio State University Wexner Medical Center between February and July 2021 (Table 1). The home visit program has been operating since 2016. In 2022, it had 30 enrolled patients. Patients were referred for home visits by their treating hematologist if they posed management

Table 1
Participant demographic characteristics (n = 11).

Variable	N (%)
Age (average years, SD)	42.82 (SD 13.93)
Gender	
Male	27.27 (%)
Female	72.72 (%)
Ethnicity	
Not Hispanic	100 (%)
Hispanic	0 (%)
Race	
African American	100 (%)
Other	0 (%)
Health insurance	
No	0 (%)
Yes	100 (%)

challenges due to high complexity and/or poor adherence to scheduled visits and treatment, and if they lacked an established primary care provider. Patients who are referred to the home visit program exhibit disproportionately high levels of emergency medical service use and receive moderate to high doses of chronic opioids for pain management. Thus, they are not representative of the sickle cell population as a whole. However, they are able to provide insight into the distress experiences of persons with sickle cell who have severe chronic pain. At the time of their study participation, all participants were living independently with no impairment in instrumental activities of daily living and no diagnosis of cognitive impairment.

The director of the home visit program (AS) and a team member (MW) discussed the study with 17 persons with sickle cell enrolled in the home visit program participants. Eleven agreed to participate. MW scheduled one-on-one interviews with each participant between March and July 2021, and conducted verbal informed consent with a waiver of written consent with each participant prior to interview start.

2.2. Data collection

A single team member (MW) conducted all interviews using the interview guide. Interviews were conducted remotely using Microsoft Teams software on an institutional laptop with data security features, and lasted 52 min on average. Conducting the interviews remotely allowed participants to interview from home instead of travelling to a central clinical site. Participants were invited to turn their cameras off. All interviews were recorded electronically using the interviewer's institutional Teams account. At the completion of the interview, Teams automatically uploaded the audio-recording of each interview to the interviewer's institutional cloud account as an MP4 file. Two team members (JC and a research coordinator) transcribed each MP4 file into a Word document and uploaded the transcription into a secure Teams channel that was established for the study.

2.3. Data analysis

Two team members (MW and a research coordinator) separately created interview memos after reviewing the transcripts and appended these to the bottom of each transcript. These memos were the first step in data analysis. They also became a valuable reference for other team members, who contributed to qualitative analysis but who had not conducted or transcribed the interviews.

The team used a deductive-dominant thematic analytical approach (Bingham & Witkowsky, 2022; Skillman et al., 2019). To achieve different study objectives, we conducted two cycles of analysis of the eleven transcripts with two different codebooks. Consistent with analytical approaches to qualitative research, our approach was iterative. In our first cycle of analysis, we analyzed the interview data for desired design and content elements for a future mindfulness mobile phone application, along with distress experiences and management strategies. We prioritized informing the content and design of the future mobile application because building this tool is a multi-year endeavor. The first cycle of analysis made full use of our interview data and the range of questions in the interview guide. While this first cycle of analysis was underway, we began a second cycle of analysis of the interview data, focusing solely on distress experiences and management approaches of participants. We excluded participants' discussions of experience with mobile applications and social media, along with their design and content preferences and needs because these data were relevant only to informing the content and design of the future mobile application. Conducting two cycles of analysis was necessary to achieve different study objectives (informing the content and design of the future mobile application and identifying sources of distress and management strategies). It was also crucial to informing the team's familiarity with the transcripts and participants' distress experiences. Findings from the first cycle of analysis will be published in a future manuscript and are informing the future mobile application development. Findings from the second cycle of analysis are presented in this manuscript.

For the first cycle of analysis, two team members (JEC and BE) created Codebook #1 in Word to identify desired application design and content elements. They imported Codebook #1 into NVivo Plus qualitative software for Windows (QSR International Pty Ltd, Released June 26, 2018), coded all transcripts to identify priorities for participants, and used content analysis to summarize priorities for the team. The team used these findings to develop the content for the mindfulness phone application.

For the second cycle of analysis, which directly informed this manuscript, JEC and BE independently reviewed each transcript in Word and marked areas of emphasis in each interview related to distress. They each manually coded all transcripts in person using colored Post-it notes, compared their coding decisions, and came to consensus on significant categories related to distress. They developed Codebook #2 in Word by using interview guide domains 1–5 and 7 as parent codes and modified these using the categories identified during manual coding. They developed definitions and inclusion and exclusion criteria for each parent and child code, imported Codebook #2 into NVivo qualitative software, and tested Codebook #2 by independently coding the same two interviews and assessing for coding disagreements and discrepancies. After revising the codebook several times through extensive discussion, they finalized Codebook #2 and coded the eleven transcripts. From these coded transcripts, they generated eight categories of distress.

To increase confidence in our findings from this meaning-centered approach, the team conducted another round of deductive-inductive analysis with additional team members. This time, four team members (JEC, BE, AS, and JC) reviewed all eleven transcripts. Each team member independently labelled passages from transcripts with de novo codes and summarized these passages for meaning related to each of the eight categories of distress. The four team members met four times to review and discuss their codes and interpretations, and to come to consensus. In the final stage, JEC and BE reviewed the coding they generated using NVivo Codebook #2, and the de novo codes and summaries generated by the four team members, to identify a conceptual focus for the manuscript with four themes. The entire team approved this focus and these themes.

3. Results

At the start of interviews, nearly all [n = 10] participants defined distress as pain and discomfort:

> When I think about distress, I think about discomfort. Right? It doesn't necessarily have to be to the extent of having a crisis, but any kind of pain or any kind of symptom that will make me feel out of the ordinary from what I would on a day-to-day basis. [Participant 13]

As interviews progressed, participants identified additional sources of distress, such as fears about addition to pain medications and interruptions to daily life. They also described the impacts of distress on them and their management approaches to sources of distress.

From the interviews, one source of distress stood out as the most disruptive and difficult to manage: anticipating and undergoing clinical encounters in the emergency department (ED) and intermediate care center (ICC) settings (hereafter "acute care settings") to manage pain flares. Their descriptions of seeking pain relief in acute care settings were characterized by anger, frustration, resentment, bewilderment, and resignation. We focused our analysis on seeking pain treatment in acute care settings as a source of distress because it had not been investigated. Although we did not collect frequency data regarding the number of times patients experienced distress from acute encounters, participants reported in their interviews that this was the most common source of distress.

After coding the interviews, we identified four themes in participants' narratives about distress related to acute care clinical encounters: (1) Pain has performative features, (2) Stigma and racism surround care, (3) Sickle cell is a neglected disease, and (4) Participants lack control over their pain management plan. We review these findings, provide quotes to illustrate each theme, and include participants' recommendations to providers.

3.1. Theme 1: Pain has performative features

Participants characterized the distress they experience seeking pain treatment in acute clinical settings as existential. One described waiting for treatment during a sickle cell flare as a threat to health and life:

> There are some of us that are perishing in emergency rooms because they don't think [that] us crying out for help and saying we need to see a doctor when we need to see a doctor is of importance. They don't understand that the longer we wait, the more damage that crisis can do not only to the part that is hurting us, but our whole entire body. [Participant 5]

The sole participant who did not define distress as pain initially described distress as having to wait in the emergency setting:

> In this context, the word "distress" is "hospital". Not just like the building, but literally being in the hospital. The room is bad enough, but if you really want to talk about distress, then we are going to have to talk about the emergency department and the typical wait, that, unless you really time it and schedule it, and if you have enough history with going to the ER to know the right time to get in there. [Participant 8]

The need for pain relief is paramount, and waiting to be seen or treated is associated with distress. But even while being seen by providers, participants experienced obstacles to obtaining pain relief. Narrating their pain is not always sufficient, they said; providers expect them to perform their pain and distress to receive treatment:

> I feel like I have to perform or show that I'm in the pain. Which is really just to make them feel comfortable about treating me even though they are supposed to, no matter what. [Participant 3]

Participants' comportment and presentation may be contrary to provider expectations of how a patient in pain should look and sound:

> That's why sometimes when doctors do an exam and they assume because we're not balled up, freaking crying and screaming our hearts, they don't think that we're in pain. [Participant 5]

Participants described provider skepticism of their pain or that they are undergoing a sickling crisis when their appearance does not conform to expectations of how a patient in pain should appear and behave:

> Sometimes they ask me, "Well, how does the pain feel? You don't look like you have sickle cell." Well, how am I supposed to look? [Participant 4]

Some participants recognized the importance of performing pain to increase the likelihood being treated promptly. For example, this participant's mother stressed the importance of demonstrating her pain in the acute care setting:

> [My mother] was like, "Okay, when we go in here you need to show them you are in a lot of pain. Because if you don't, we are going to be here forever. You're going to suffer." [Participant 3]

For some, pain performance begins before arriving at the acute care setting. Participants described dressing the part of a patient in pain before leaving home:

> You'd be surprised at how many of us sickle cell patients have to undress before going to the hospital. By that I mean, taking eyelashes off if possible, put pajamas on, you know. You don't want to go to the hospital looking a certain type of way and they look at you like, "Oh you're not sick, and we're not going to treat you." [Participant 2]

But it was clear that not all can perform their pain for providers:

> A lot of people can cry on cue. I can't do that. I don't know how to act. I can't fake [it]. [Participant 2]

Performing and expressing pain can worsen the experience of it. For these participants, minimizing pain expression is an important strategy to manage their pain and distress while waiting for treatment in the acute care setting. Some participants manage their pain and distress by minimizing facial and bodily expressions, even while aware that this may reduce their credibility to providers:

> I feel like I had to like moan louder and move around a lot. And that was never good for me. Moving around a lot and expressing that I'm in pain feeds into the pain more. It gives it more attention, therefore I feel it more. [Participant 3]

Participants asked that providers look beyond the presence or absence of facial and bodily expressions to assess their pain:

> Even though I can carry on a conversation like everything seems normal when I'm in a crisis, it doesn't mean that I'm not in a crisis. Look deeper than that. Don't focus on just the outside because it is not the outside that is hurting. It's the inside. [Participant 5]

3.2. Theme 2: Stigma and racism surround care

The challenges associated with being a person of color seeking treatment for acute pain were woven into participants' narratives about distress and the acute care setting. They described how providers perceive them as drug-seeking because they are black/African-American. Participants perceived this stigma curtailed their ability to advocate for themselves by asking for the medication they need:

> I need more pain medication then what I've been given. But I can't ask for it. Because I'm black, I'll be seen as drug seeking. I've had several comments made to me before that suggest that a doctor or a nurse thought that I was drug-seeking because of my race and strictly because I had sickle cell. I feel like my nurses and doctors don't trust me. [Participant 3]

> I feel like a lot of the doctors and nurses are racist. They think that all of us drug seekers, you know? Because they deal with so many patients that come in wanting drugs, we all just fall right in line. Being African-American with pain is just double-trouble. [Participant 2]

Participants recommended providers and hospital staff develop awareness of their implicit bias and professional oaths to provide equitable care:

> Just because I have sickle cell disease and I'm a woman of color does not mean that I am there to utilize the hospital system in anyway shape or form. Don't judge me based off of my skin color. I want to have the same, equal care that a person from a different ethnicity would have. The same exact care that you would give to your loved one is the same exact care that I should be able to have. You took that oath of being a healthcare provider. There's definitely ethics that go along with being a provider. You are there to serve. [Participant 13]

Another expressed the desire for providers to listen to them and treat them as credible experts on their own pain:

> When I say how much pain I'm in, I'm telling the truth. You need to be open to the idea that you're wrong about how much pain medication I actually need. And be willing to communicate with me instead of just tell me what you're going to do. It needs to be a dialogue. [Participant 3]

3.3. Theme 3: Sickle cell is a neglected disease

Participants characterized sickle cell as a neglected disease, in two senses. They felt it was neglected in medical education and clinical training. For example, they observed that providers seem unprepared to manage persons with sickle cell appropriately and felt they received less attention and pain relief compared to patients with cancers or cystic fibrosis:

> They don't really know what sickle cell does to our bodies. Not walking a mile in our shoes to understand the pain is unbearable and we can't handle it. I've had my fair share of going off on nurses and doctors that doesn't get it. They say they know about sickle cell. Obviously, you haven't done your research deeply enough to really understand it. [Participant 11]

Some expressed astonishment that some providers in the acute care setting are unfamiliar with sickle cell:

> [The] Caucasian lady was like, "I've never heard of sickle cell." And I was like, what the hell? How are you a nurse in the ER and you never heard of a sickle cell? [Participant 14]

One participant attributed providers' lack of awareness of sickle cell to it being considered a "black" disease:

> Like I've said, it's because it's a black disease that people just don't know about it, and it's not talked about. [Participant 12]

They expressed the desire for providers to develop a better clinical understanding of sickle cell in their undergraduate medical education and residency training. One participant called for increased representation of physicians in acute care settings who are themselves persons of color and have had extensive professional experience with sickle cell:

> We need black doctors that actually know about sickle cell and been deeply studying it for years. I'm talking about doctors that are from Africa, doctors from Honduras, Jamaica, Haiti. [Participant 11]

Participants also characterized sickle cell as a disease that was neglected in research and treatment investment compared to other diseases that cause pain but that do not primarily affect persons of color. They observed that patients with cancers and cystic fibrosis appear to benefit from significantly more funding dollars, greater public attention and sympathy, and preferential clinical treatment:

> They mention their cancer patients a lot. It's annoying because I have to go to a hospital that's mainly dedicated to cancer. I'm always in spaces that say cancer on the door. [Participant 3]

One participant called out what they perceived as disparities in dispensing pain medication between sickle cell and cancer patients:

> They were going to stop providing monthly prescriptions of pain medications for pretty much everyone unless you are a cancer patient. If you are not a cancer patient, then you are limited to getting a week's worth of pain medication. [Participant 7]

3.4. Theme 4: Participants lack control over their pain management plan

Participants reported that, once they were seen by providers in acute care settings, providers might change their medications or pain plan without consulting them:

> I have no choice but to come to the emergency department and get treatment. But at the same time, it's not fair that doctors are taking our medicine away from us and giving us something that we don't even need or never had or never tried before. [They're] changing our pain plan without consulting with us first. [Participant 11]

They expressed frustration that providers do not see persons as sickle cell as experts on their own bodies and pain:

> When we're in the hospital, many times I tried to tell doctors like this will work, this is what I need, let's get this going so I can get out of here. I know my body. If I'm there, I'm there for a reason. I hate the hospital. I feel like it's jail. [Participant 12]

> I've always just been told this is what you're getting and be happy with it. And it's even worse now because of the opioid crisis. I'd like them to know that my pain may not fit the descriptions you have. [Participant 3]

Yet some hesitate to advocate for themselves if the acute care setting is not their usual place of treatment:

> [The provider said,] "I'm giving you [dosage of pain medication] and you might be sleepy and if you pass out, just let me know." And I was like, "Woman, I've been on pain medicine my entire life. That's going to barely touch it or give me any relief, because I will sometimes get [higher dosage of pain medication]." But I didn't go there and tell her that because I wasn't at my home hospital. [Participant 14]

Participants instead may ask family or friends to advocate to providers on their behalf:

> I have been in a lot of situations where I couldn't even get pain medications until somebody showed up to the hospital for me. [Participant 13]

They reported that providers have threatened to take away their pain medicine if they express the desire to pursue alternative pain management approaches:

> So [the provider] tells me "Oh well, we don't believe in medical marijuana." You mean *you* don't believe in medical marijuana, but I do. I believe that is something that is provided for us to have. And I get hit with the, "Well, if you're doing medical marijuana then we're taking your pain meds from you." [Participant 11]

As a recommendation, participants ask providers to recognize they know their bodies and to act on the information they share with them:

> Listen to us when we say, because we know our bodies, if we don't have ports, we know the best thing to do is to start an IV. Just listen to us. That's all we want: for you to listen to us and fully understand us because we're coming to you for help. [Participant 5]

4. Discussion

Seeking pain treatment in acute care settings is a profound source of distress for persons with sickle cell. This distress is related to acute pain itself. It is also related to postponing seeking treatment, waiting for treatment in the acute care setting, past experiences of mistreatment by

providers, and lack of control over pain management in these settings.

Participants in our study, all of whom are persons of color, drew attention to the intersectional stigma of racial bias, pain, and sickle cell disease in these settings (Khatri et al., 2023). Their distress often begins with anticipating an acute care visit and may extend through the visit itself. They described entering the acute care setting for pain treatment with anger, frustration, resentment, bewilderment, and resignation.

This is a startling finding, because it tells us that a potentially powerful and important act of self-care for persons with sickle cell—seeking clinical care for acute pain that is unmanageable outside of a clinical setting—is a complex source of distress and challenging to manage. Participants readily identified multiple strategies to manage other sources of distress in their daily lives (e.g., prayer, singing, exercise, distraction, taking hot baths), and they communicated satisfaction, even a sense of mastery, in describing these. However, participants identified only four strategies to manage distress associated with seeking acute pain treatment or being in the acute care setting: avoiding or postponing care; enacting or withholding pain performance; asking a family member to advocate for their treatment; and distracting themselves from their pain. They expressed frustration and impotence when describing these strategies. Pain management strategies deployed by acute care settings that are not effectively working for patients may exacerbate their experiences of chronic social defeat and increase hesitation when seeking care in acute care settings. An important inference is that by prioritizing the autonomy of persons with sickle cell and their shared decision making in the treatment of their pain in acute care settings, it may be possible to reduce their distress and associated pain.

Our findings, that participants have few management strategies for distress related to the acute setting, and that they do not express the same degree of satisfaction in employing these strategies as with other strategies to manage distress in daily life, do not suggest an absence of creativity or resourcefulness in self-management for this population. Rather, these findings point to the power asymmetries that persons with sickle cell and their families experience while seeking pain relief in acute care settings. This is a source of distress that is not well-captured by psychosocial models of distress, which seek to measure distress but not to explore the complex systems or social norms that maintain it.

Seeking pain treatment in an acute care setting means being seen by providers who may be unfamiliar with them as patients or with the pain management plan developed with their regular providers. Our study found that persons who have chronic pain with sickle cell are aware of the care they seek from providers in acute care settings, yet they have few strategies to meet their care needs. Participants mentioned the stigma by providers when they ask for pain medication, a common finding in qualitative research with this population. They recommended that providers listen to persons with sickle cell and have a dialogue with them about pain management. Given that persons with sickle cell place high value on honest and open communication with providers, and they desire a shared pain management plan, the lack of familiarity of acute care providers with them and their care has profound implications for their well-being.

Participants compared the quality of treatment for acute pain flares unfavorably to acute pain treatment for persons with cancers and cystic fibrosis. Their comparison of these diseases underscores a sense of abandonment as a patient population by researchers and the medical profession, and a perception that racial bias in acute care settings adversely impacts pain treatment. It is clear that sickle cell disease receives significantly less research investment than diseases that impact similar-sized populations, such as cystic fibrosis (Farooq et al., 2020). Compared to cystic fibrosis, sickle cell is a neglected disease, and the reasons for this neglect are structural (Bahr & Song, 2015).

Our analysis introduces the notion of pain performativity in the acute care setting. Participants told us that validation of their pain and its treatment by providers may depend on the "correct" performance of a set of culturally accepted norms of facial and bodily pain expression that providers recognize. Participants described not simply arriving in the acute care setting while in pain, aware of the complications they might be having. They also anticipated a long wait to be seen and prepared themselves to offer a stylized presentation to providers as a patient in pain. This is evident with the participant who "dressed for the occasion" at her mother's insistence by not grooming herself for the visit. This strategy has been identified in a qualitative study with persons with sickle cell who described altering their appearance or presentation to providers in the acute care setting in response to the opioid-related stigma they encountered (Abu Al Hamayel et al., 2021).

One way to describe this approach of pain performance is "managing the other"—that is, managing the perceptions of treating providers to achieve desired outcomes (Nwora et al., 2024). A mixed-methods study with young adults aged 19–25 years with sickle cell found that participants adopt a range of management strategies that were associated with the theme of "being a good patient" and included both "managing the self" and "managing the other" (Nwora et al., 2024). In this study, as in ours, participants stressed the importance of coming to the acute care setting prepared to obtain effective treatment. But rather than trying to "look the part" of patients in distress or pain, the young adults in this study emphasized advocating for themselves in acute care settings by telling providers what treatments they need ("managing the self"), as well as empathizing with clinicians ("managing the other"). Our participant population, which is older, did not state that advocating for themselves was an effective strategy. More often, they associated it with defeat, or with the need to have someone advocate for them. Additionally, their encounters in acute care settings were characterized by adversity and tension. They did not identify empathizing with providers as a management strategy for their clinical encounters. Researchers may wish to investigate which variables, such as age, are associated with the adoption of specific strategies to manage acute care encounters and identify outcomes of these strategies from the perspective of patients and providers.

Our findings suggest that a "failure to perform" in ways that persuade providers of the legitimacy of their pain has consequences. Two consequences that participants identified are provider skepticism of a pain crisis and difficulty of the participant in obtaining adequate pain treatment. Because participants have been greeted with skepticism about their pain, labelled as drug-seeking, and enduring long waits, most reported that they avoid or postpone seeking treatment for acute pain as long as possible. This avoidance inevitably makes clinical management of their crisis and pain more challenging.

It is important to acknowledge that providers may experience frustration, impotence, and moral distress from their inability to effectively manage acute pain in persons with sickle cell, and this may contribute to the distress of persons with sickle in acute care settings. Many persons with sickle cell are managed with chronic opioids continuously from a young age, resulting in tolerance to opioids and a requirement for high doses to achieve effective pain relief. The conflict between the high dose requirements of sickle cell patients and the strong social mandate to reduce opioid prescribing may be a source of considerable discomfort and conflict to providers. This may contribute to viewing persons with sickle cell as difficult. Reliance on the hematologic model of sickle cell pain may compound providers' challenges in diagnosing and treating acute pain effectively (Childerhose et al., 2023).

Existing investigation of pain communication in clinical settings offers some opportunities for investigation and intervention development. Findings include that both providers and patients may minimize pain sensations, effective communication is relational (built by patient and provider over time), and certain tools, such as pain cards or photos, may foster a collaborative approach to discussing pain (Galatolo & Fasulo, 2022; Semino et al., 2017; Weatherall et al., 2021). Photovoice, for example, has been used to support persons with sickle cell in communicating their distress and educating providers (Young et al., 2020). However, there has been little investigation of how patients make decisions about performing pain and withholding pain cues, and which performative norms for pain providers recognize and validate in acute

care settings. There may be value in adopting a dramaturgical approach to understand the miscues between persons with sickle cell and providers in these settings (Goffman, 2002; Ramsey et al., 2023).

For the ongoing study of distress in persons with sickle cell, there is merit in considering how the acute setting and opioid deprescribing guidelines may create a nexus of distress for persons with sickle cell, their families, and providers. This requires looking beyond the patient as an individual who is responsible for their own management of distress and asking how their interactions with their broader pain management context shape their distress and their experience of pain. By focusing on interventions that address these dynamics, it may be possible to transform the acute care clinical encounter from a profound source of distress for persons with sickle cell into a source of relief and support.

5. Conclusion

Seeking pain treatment in acute care settings is a profound and complex source of distress for persons with sickle cell. A decision to seek pain treatment may, paradoxically, potentiate pain and erode trust. Researchers may wish to consider how acute care settings and practices foster distress. We believe there are opportunities to investigate how providers interpret and validate specific pain presentations in acute care settings, and to identify additional strategies to help this population manage distress fostered by seeking acute care using mindfulness-based educational tools. To support persons with sickle cell, providers may wish to adopt recommendations from study participants: treat them as credible experts on their own pain, look beyond the presence of absence of pain performance, develop awareness of implicit bias, improve clinical understanding of sickle cell, and act on the treatment information that patients share. Adopting these practices may help providers achieve an outcome that patients also seek: the effective treatment of acute pain.

Study limitations

This study was the first step in engaging persons with sickle cell who are enrolled in a home visit program. The purpose of the study was to inform the development of a future mobile phone application for use by persons with sickle cell. We will separately publish quantitative data on the content, design, and effectiveness of the future mobile application to help patients reframe their pain experiences.

The design for this study objective was meaning-centered and qualitative. Consequently, investigators did not measure the distress or pain experiences of study participants. Our study sample was small and not representative of the wide range of disease severity. We used convenience sampling to recruit home visit participants who were treatment-resistant, with the hope we would gain insight into the concerns and needs of the most severely impacted patients. Because we used convenience sampling, we did not consider sex and gender in our sampling approach. These approaches limit the generalizability of our findings.

Declarations

None.

Ethics approval and consent to participate

All procedures were performed in compliance with relevant laws and institutional guidelines and have been approved by the appropriate institutional committee(s). The study, entitled Development of a Mobile Application to Train Adults with Sickle Cell Disease in Mindfulness-Based Pain Management, was approved by The Ohio State University Institutional Review Board in February 2021 (2021H0032). Verbal informed consent with a waiver of written consent was obtained from all study participants.

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CRediT authorship contribution statement

Janet E. Childerhose: Writing – review & editing, Writing – original draft, Supervision, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Brent Emerson:** Writing – review & editing, Writing – original draft, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Andrew Schamess:** Writing – review & editing, Writing – original draft, Formal analysis, Conceptualization. **Jacqueline Caputo:** Writing – review & editing, Writing – original draft, Conceptualization, Data curation, Formal analysis, Investigation, Methodology. **Marcus Williams:** Writing – review & editing, Project administration, Investigation, Conceptualization. **Maryanna D. Klatt:** Writing – review & editing, Visualization, Validation, Supervision, Software, Project administration, Methodology, Investigation, Funding acquisition, Data curation, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

Abdallah, K., Buscetta, A., Cooper, K., Byeon, J., Crouch, A., Pink, S., et al. (2020). Emergency department utilization for patients living with sickle cell disease: Psychosocial predictors of health care behaviors. *Annals of Emergency Medicine, 76*, S56–S63. https://doi.org/10.1016/j.annemergmed.2020.08.018

Abu Al Hamayel, N., Waldfogel, J. M., Hannum, S. M., Brodsky, R. A., Bolanos-Meade, J., Gamper, C. J., et al. (2021). Pain experiences of adults with sickle cell disease and hematopoietic stem cell transplantation: A qualitative study. *Pain Medicine, 22*, 1753–1759. https://doi.org/10.1093/pm/pnaa464

Adegbola, M. A., Barnes, D. M., Opollo, J. G., Herr, K., Gray, J., & McCarthy, A. M. (2012). Voices of adults living with sickle cell disease pain. *Journal of National Black Nurses' Association, 23*, 16–23.

Aisiku, I. P., Penberthy, L. T., Smith, W. R., Bovbjerg, V. E., McClish, D. K., Levenson, J. L., et al. (2007). Patient satisfaction in specialized versus nonspecialized adult sickle cell care centers: The PiSCES study. *Journal of the National Medical Association, 99*, 886–890. https://www.ncbi.nlm.nih.gov/pubmed/17722665.

Badawy, S. M., Cronin, R. M., Hankins, J., Crosby, L., DeBaun, M., Thompson, A. A., et al. (2018). Patient-centered ehealth interventions for children, adolescents, and adults with sickle cell disease: Systematic review. *Journal of Medical Internet Research, 20*, Article e10940. https://doi.org/10.2196/10940

Bahr, N. C., & Song, J. (2015). The effect of structural violence on patients with sickle cell disease. *Journal of Health Care for the Poor and Underserved, 26*, 648–661. https://doi.org/10.1353/hpu.2015.0094

Biltz, R. G., Sawicki, C. M., Sheridan, J. F., & Godbout, J. P. (2022). The neuroimmunology of social-stress-induced sensitization. *Nature Immunology, 23*, 1527–1535. https://doi.org/10.1038/s41590-022-01321-z

Bingham, A. J., & Witkowsky, P. (2022). Deductive and inductive approaches to qualitative data analysis. In C. Vanover, P. Mihas, & J. Saldaña (Eds.), *Analyzing and interpreting qualitative data: After the interview* (pp. 133–146). Thousand Oaks, CA: SAGE Publications.

Booker, M. J., Blethyn, K. L., Wright, C. J., & Greenfield, S. M. (2006). Pain management in sickle cell disease. *Chronic Illness, 2*, 39–50. https://doi.org/10.1177/17423953060020011101

Brintz, C. E., Roth, I., Faurot, K., Rao, S., & Gaylord, S. A. (2020). Feasibility and acceptability of an abbreviated, four-week mindfulness program for chronic pain management. *Pain Medicine, 21*, 2799–2810. https://doi.org/10.1093/pm/pnaa208

Bulgin, D., Tanabe, P., & Jenerette, C. (2018). Stigma of sickle cell disease: A systematic review. *Issues in Mental Health Nursing, 39*, 675–686. https://doi.org/10.1080/01612840.2018.1443530

Campbell, C. M., Moscou-Jackson, G., Carroll, C. P., Kiley, K., Haywood, C., Jr., Lanzkron, S., et al. (2016). An evaluation of central sensitization in patients with sickle cell disease. *The Journal of Pain, 17*, 617–627. https://doi.org/10.1016/j.jpain.2016.01.475

Carroll, C. P., Haywood, C., Jr., Fagan, P., & Lanzkron, S. (2009). The course and correlates of high hospital utilization in sickle cell disease: Evidence from a large, urban Medicaid managed care organization. *American Journal of Hematology, 84*, 666–670. https://doi.org/10.1002/ajh.21515

Carroll, C. P., Haywood, C., Jr., & Lanzkron, S. (2011). Prediction of onset and course of high hospital utilization in sickle cell disease. *Journal of Hospital Medicine, 6*, 248–255. https://doi.org/10.1002/jhm.850

Childerhose, J. E., Cronin, R. M., Klatt, M. D., & Schamess, A. (2023). Treating chronic pain in sickle cell disease - the need for a biopsychosocial model. *New England Journal of Medicine, 388*, 1349–1351. https://doi.org/10.1056/NEJMp2301143

Citero Vde, A., Levenson, J. L., McClish, D. K., Bovbjerg, V. E., Cole, P. L., Dahman, B. A., et al. (2007). The role of catastrophizing in sickle cell disease–the PiSCES project. *Pain, 133*, 39–46. https://doi.org/10.1016/j.pain.2007.02.008

Coleman, B., Ellis-Caird, H., McGowan, J., & Benjamin, M. J. (2016). How sickle cell disease patients experience, understand and explain their pain: An interpretative phenomenological analysis study. *British Journal of Health Psychology, 21*, 190–203. https://doi.org/10.1111/bjhp.12157

Crego, N., Masese, R., Bonnabeau, E., Douglas, C., Rains, G., Shah, N., et al. (2021). Patient perspectives of sickle cell management in the emergency department. *Critical Care Nursing Quarterly, 44*, 160–174. https://doi.org/10.1097/CNQ.0000000000000350

Creswell, J. D. (2017). Mindfulness interventions. *Annual Review of Psychology, 68*, 491–516. https://doi.org/10.1146/annurev-psych-042716-051139

Dampier, C., Barry, V., Gross, H. E., Lui, Y., Thornburg, C. D., DeWalt, D. A., et al. (2016a). Initial evaluation of the pediatric PROMIS(R) health domains in children and adolescents with sickle cell disease. *Pediatric Blood and Cancer, 63*, 1031–1037. https://doi.org/10.1002/pbc.25944

Dampier, C., Jaeger, B., Gross, H. E., Barry, V., Edwards, L., Lui, Y., et al. (2016b). Responsiveness of PROMIS(R) pediatric measures to hospitalizations for sickle pain and subsequent recovery. *Pediatric Blood and Cancer, 63*, 1038–1045. https://doi.org/10.1002/pbc.25931

Edwards, C. L., Green, M., Wellington, C. C., Muhammad, M., Wood, M., Feliu, M., et al. (2009). Depression, suicidal ideation, and attempts in black patients with sickle cell disease. *Journal of the National Medical Association, 101*, 1090–1095. https://doi.org/10.1016/s0027-9684(15)31103-2

Edwards, C. L., Killough, A., Wood, M., Doyle, T., Feliu, M., Barker, C. S., et al. (2014). Emotional reactions to pain predict psychological distress in adult patients with sickle cell disease (SCD). *The International Journal of Psychiatry in Medicine, 47*, 1–16. https://doi.org/10.2190/PM.47.1.a

Elahi, N., Tahery, N., Ahmadi, F., & Rostami, S. (2017). Lived experienced of patients with sickle cell disease anemia about disease management (a qualitative research). *Annals of Tropical Medicine and Public Health, 10*, 1238–1242. https://doi.org/10.4103/Atmph.Atmph_239_17

Ezenwa, M. O., Yao, Y., Molokie, R. E., Wang, Z. J., Mandernach, M. W., Suarez, M. L., et al. (2017). Coping with pain in the face of healthcare injustice in patients with sickle cell disease. *Journal of Immigrant and Minority Health, 19*, 1449–1456. https://doi.org/10.1007/s10903-016-0432-0

Farooq, F., Mogayzel, P. J., Lanzkron, S., Haywood, C., & Strouse, J. J. (2020). Comparison of US federal and foundation funding of research for sickle cell disease and cystic fibrosis and factors associated with research productivity. *JAMA Network Open, 3*, Article e201737. https://doi.org/10.1001/jamanetworkopen.2020.1737

Galatolo, R., & Fasulo, A. (2022). Talking down pain in the prosthesis clinic: The emergence of a local preference. *Research on Language and Social Interaction, 55*, 101–121. https://doi.org/10.1080/08351813.2022.2026172

Gilbertson, R. M., & Klatt, M. D. (2017). Mindfulness in motion for people with multiple sclerosis: A feasibility study. *Int J MS Care, 19*, 225–231. https://doi.org/10.7224/1537-2073.2015-095

Goffman, E. (2002). *The Presentation of Self in everyday life. 1959*. Garden City, NY: Doubleday & Company.

Grossman, P., Niemann, L., Schmidt, S., & Walach, H. (2004). Mindfulness-based stress reduction and health benefits. A meta-analysis. *Journal of Psychosomatic Research, 57*, 35–43. https://doi.org/10.1016/S0022-3999(03)00573-7

Hays, R. D., Spritzer, K. L., Schalet, B. D., & Cella, D. (2018). PROMIS(R)-29 v2.0 profile physical and mental health summary scores. *Quality of Life Research, 27*, 1885–1891. https://doi.org/10.1007/s11136-018-1842-3

Haywood, C., Jr., Diener-West, M., Strouse, J., Carroll, C. P., Bediako, S., Lanzkron, S., et al. (2014). Perceived discrimination in health care is associated with a greater burden of pain in sickle cell disease. *Journal of Pain and Symptom Management, 48*, 934–943. https://doi.org/10.1016/j.jpainsymman.2014.02.002

Haywood, C., Jr., Lanzkron, S., Ratanawongsa, N., Bediako, S. M., Lattimer, L., Powe, N. R., et al. (2010). The association of provider communication with trust among adults with sickle cell disease. *Journal of General Internal Medicine, 25*, 543–548. https://doi.org/10.1007/s11606-009-1247-7

Jacob, E. (2001). The pain experience of patients with sickle cell anemia. *Pain Management Nursing, 2*, 74–83. https://doi.org/10.1053/jpmn.2001.26119

Jenerette, C. M., Brewer, C. A., & Ataga, K. I. (2014). Care seeking for pain in young adults with sickle cell disease. *Pain Management Nursing, 15*, 324–330. https://doi.org/10.1016/j.pmn.2012.10.007

Jonassaint, C. R., Shah, N., Jonassaint, J., & De Castro, L. (2015). Usability and feasibility of an mhealth intervention for monitoring and managing pain symptoms in sickle cell disease: The sickle cell disease mobile application to record symptoms via technology (SMART). *Hemoglobin, 39*, 162–168. https://doi.org/10.3109/03630269.2015.1025141

Kabat-Zinn, J. (2005). *Wherever you go, there you are: Mindfulness meditation in everyday life*. New York: Hyperion.

Kabat-Zinn, J., Lipworth, L., & Burney, R. (1985). The clinical use of mindfulness meditation for the self-regulation of chronic pain. *Journal of Behavioral Medicine, 8*, 163–190. https://doi.org/10.1007/BF00845519

Karafin, M. S., Chen, G., Wandersee, N. J., Brandow, A. M., Hurley, R. W., Simpson, P., et al. (2019). Chronic pain in adults with sickle cell disease is associated with alterations in functional connectivity of the brain. *PLoS One, 14*, Article e0216994. https://doi.org/10.1371/journal.pone.0216994

Keller, S., Yang, M., Treadwell, M. J., & Hassell, K. L. (2017). Sensitivity of alternative measures of functioning and wellbeing for adults with sickle cell disease: Comparison of PROMIS(R) to ASCQ-Me. *Health and Quality of Life Outcomes, 15*, 117. https://doi.org/10.1186/s12955-017-0661-5

Keller, S. D., Yang, M., Treadwell, M. J., Werner, E. M., & Hassell, K. L. (2014). Patient reports of health outcome for adults living with sickle cell disease: Development and testing of the ASCQ-Me item banks. *Health and Quality of Life Outcomes, 12*, 125. https://doi.org/10.1186/s12955-014-0125-0

Khatri, U. G., Nguemeni Tiako, M. J., Gebreyesus, A., Reid, A., Jacoby, S. F., & South, E. C. (2023). "A lack of empathy:" A qualitative study of Black people seeking treatment for opioid use disorder. *SSM - Qualitative Research in Health, 4*, Article 100298. https://doi.org/10.1016/j.ssmqr.2023.100298

Knisely, M. R., Tanabe, P. J., Yang, Q., Masese, R., Jiang, M., & Shah, N. R. (2021). Severe pain profiles and associated sociodemographic and clinical characteristics in individuals with sickle cell disease. *The Clinical Journal of Pain, 37*, 669–677. https://doi.org/10.1097/AJP.0000000000000957

Lanzkron, S., Little, J., Field, J., Shows, J. R., Wang, H., Seufert, R., et al. (2018). Increased acute care utilization in a prospective cohort of adults with sickle cell disease. *Blood Adv, 2*, 2412–2417. https://doi.org/10.1182/bloodadvances.2018018382

Levenson, J. L., McClish, D. K., Dahman, B. A., Penberthy, L. T., Bovbjerg, V. E., Aisiku, I. P., et al. (2007). Alcohol abuse in sickle cell disease: The PiSCES Project. *American Journal on Addictions, 16*, 383–388. https://doi.org/10.1080/10550490701525434

Liederman, Z., Quartey, N. K., Ward, R., & Papadakos, J. (2021). Exploration of barriers and facilitators to optimal emergency department care of sickle cell disease: Opportunities for patient-physician partnerships to improve care. *Hemoglobin, 45*, 13–19. https://doi.org/10.1080/03630269.2020.1859383

Matthie, N., & Jenerette, C. (2017). Understanding the self-management practices of young adults with sickle cell disease. *J Sick Cell Dis Hemoglobinopathies, 2017*, 76–87.

Matthie, N., Jenerette, C., Gibson, A., Paul, S., Higgins, M., & Krishnamurti, L. (2020). Prevalence and predictors of chronic pain intensity and disability among adults with sickle cell disease. *Health Psychology Open, 7*, Article 2055102920917250. https://doi.org/10.1177/2055102920917250

Matthie, N., Jenerette, C., & McMillan, S. (2015). Role of self-care in sickle cell disease. *Pain Management Nursing, 16*, 257–266. https://doi.org/10.1016/j.pmn.2014.07.003

Matthie, N., Ross, D., Sinha, C., Khemani, K., Bakshi, N., & Krishnamurti, L. (2019). A qualitative study of chronic pain and self-management in adults with sickle cell disease. *Journal of the National Medical Association, 111*, 158–168. https://doi.org/10.1016/j.jnma.2018.08.001

McClish, D. K., Penberthy, L. T., Bovbjerg, V. E., Roberts, J. D., Aisiku, I. P., Levenson, J. L., et al. (2005). Health related quality of life in sickle cell patients: The PiSCES project. *Health and Quality of Life Outcomes, 3*, 50. https://doi.org/10.1186/1477-7525-3-50

McClish, D. K., Smith, W. R., Dahman, B. A., Levenson, J. L., Roberts, J. D., Penberthy, L. T., et al. (2009). Pain site frequency and location in sickle cell disease: The PiSCES project. *Pain, 145*, 246–251. https://doi.org/10.1016/j.pain.2009.06.029

McClish, D. K., Smith, W. R., Levenson, J. L., Aisiku, I. P., Roberts, J. D., Roseff, S. D., & Bovbjerg, V. E. (2017). Comorbidity, pain, utilization, and psychosocial outcomes in older versus younger sickle cell adults: The PiSCES project. *BioMed Research International, 2017*, Article 4070547. https://doi.org/10.1155/2017/4070547

Nwora, C., Prince, E. J., Pugh, L., Weaver, M. S., & Pecker, L. H. (2024). How young adults with sickle cell disease define "being a good patient" in the adult healthcare system. *Pediatric Blood and Cancer, 71*, Article e30786. https://doi.org/10.1002/pbc.30786

Pope, J. E., Fishman, M., Chakravarthy, K., Hanes, M., Gerling, M., Heros, R., et al. (2021). A retrospective, multicenter, quantitative analysis of patients' baseline pain quality (PROMIS-29) entering into pain and spine practices in the United States (ALIGN). *Pain Ther, 10*, 539–550. https://doi.org/10.1007/s40122-021-00238-z

Porter, L. S., Gil, K. M., Sedway, J. A., Ready, J., Workman, E., & Thompson, R. J., Jr. (1998). Pain and stress in sickle cell disease: An analysis of daily pain records. *International Journal of Behavioral Medicine, 5*, 185–203. https://doi.org/10.1207/s15327558ijbm0503_1

Power-Hays, A., & McGann, P. T. (2020). When actions speak louder than words - racism and sickle cell disease. *New England Journal of Medicine, 383*, 1902–1903. https://doi.org/10.1056/NEJMp2022125

Prince, E. J., Pecker, L. H., Lanzkron, S., & Carroll, C. P. (2023). The complex association of daily opioid dose with visits for pain in sickle cell disease: Tolerance or treatment-refractory pain? *Pain Medicine, 24*, 703–712. https://doi.org/10.1093/pm/pnac187

QSR International Pty Ltd. (Released June 26, 2018). NVivo Plus Version 12.1.

Ramsey, L., O'Hara, J., Lawton, R., & Sheard, L. (2023). A glimpse behind the organisational curtain: A dramaturgical analysis exploring the ways healthcare staff engage with online patient feedback 'front' and 'backstage' at three hospital trusts in england. *Sociology of Health & Illness, 45*, 642–665. https://doi.org/10.1111/1467-9566.13607

Reeve, B. B., Hays, R. D., Bjorner, J. B., Cook, K. F., Crane, P. K., Teresi, J. A., et al. (2007). Psychometric evaluation and calibration of health-related quality of life item banks: Plans for the Patient-Reported Outcomes Measurement Information System (PROMIS). *Medical Care, 45*, S22–S31. https://doi.org/10.1097/01.mlr.0000250483.85507.04

Renedo, A., Miles, S., Chakravorty, S., Leigh, A., Telfer, P., Warner, J. O., et al. (2019). Not being heard: Barriers to high quality unplanned hospital care during young people's transition to adult services - evidence from 'this sickle cell life' research. *BMC Health Services Research, 19*, 876. https://doi.org/10.1186/s12913-019-4726-5

Robbins, M. A., McGill, L. S., Holloway, B. M., & Bediako, S. M. (2019). Sickle cell disease, more than just pain: The mediating role of psychological symptoms. *Southern Medical Journal, 112*, 253–258. https://doi.org/10.14423/SMJ.0000000000000972

Santos, L., Guimaraes, M. W., Baptista, A. F., & Sa, K. N. (2021). Impact of neuropathic pain on quality of life in adults with sickle cell disease: Observational study. *Hematol Transfus Cell Ther, 43*, 263–267. https://doi.org/10.1016/j.htct.2020.03.010

Schuman-Olivier, Z., Trombka, M., Lovas, D. A., Brewer, J. A., Vago, D. R., Gawande, R., et al. (2020). Mindfulness and behavior change. *Harvard Review of Psychiatry, 28*, 371–394. https://doi.org/10.1097/HRP.0000000000000277

Semino, E., Zakrzewska, J. M., & Williams, A. (2017). Images and the dynamics of pain consultations. *The Lancet, 389*, 1186–1187.

Shah, N., Jonassaint, J., & De Castro, L. (2014). Patients welcome the sickle cell disease mobile application to record symptoms via technology (SMART). *Hemoglobin, 38*, 99–103. https://doi.org/10.3109/03630269.2014.880716

Simmons, L. A., Williams, H., Silva, S., Keefe, F., & Tanabe, P. (2019). Acceptability and feasibility of a mindfulness-based intervention for pain catastrophizing among persons with sickle cell disease. *Pain Management Nursing, 20*, 261–269. https://doi.org/10.1016/j.pmn.2018.10.002

Sinha, C. B., Bakshi, N., Ross, D., & Krishnamurti, L. (2019). Management of chronic pain in adults living with sickle cell disease in the era of the opioid epidemic: A qualitative study. *JAMA Network Open, 2*, Article e194410. https://doi.org/10.1001/jamanetworkopen.2019.4410

Skillman, M., Cross-Barnet, C., Friedman Singer, R., Rotondo, C., Ruiz, S., & Moiduddin, A. (2019). A framework for rigorous qualitative research as a component of mixed method rapid-cycle evaluation. *Qualitative Health Research, 29*, 279–289. https://doi.org/10.1177/1049732318795675

Smith, W. R., Bovbjerg, V. E., Penberthy, L. T., McClish, D. K., Levenson, J. L., Roberts, J. D., et al. (2005). Understanding pain and improving management of sickle cell disease: The PiSCES study. *Journal of the National Medical Association, 97*, 183–193.

Smith, W. R., McClish, D. K., Levenson, J., Aisiku, I., Dahman, B., Bovbjerg, V. E., et al. (2018). Predictive ability of intermittent daily sickle cell pain assessment: The PiSCES project. *Pain Medicine, 19*, 1972–1981. https://doi.org/10.1093/pm/pnx214

Smith, W. R., Penberthy, L. T., Bovbjerg, V. E., McClish, D. K., Roberts, J. D., Dahman, B., et al. (2008). Daily assessment of pain in adults with sickle cell disease. *Annals of Internal Medicine, 148*, 94–101. https://doi.org/10.7326/0003-4819-148-2-200801150-00004

Sogutlu, A., Levenson, J. L., McClish, D. K., Rosef, S. D., & Smith, W. R. (2011). Somatic symptom burden in adults with sickle cell disease predicts pain, depression, anxiety, health care utilization, and quality of life: The PiSCES project. *Psychosomatics, 52*, 272–279. https://doi.org/10.1016/j.psym.2011.01.010

Thomas, V. J., & Taylor, L. M. (2002). The psychosocial experience of people with sickle cell disease and its impact on quality of life: Qualitative findings from focus groups. *British Journal of Health Psychology, 7*, 345–363. https://doi.org/10.1348/135910702760213724

Treadwell, M. J., Barreda, F., Kaur, K., & Gildengorin, G. (2015). Emotional distress, barriers to care, and health-related quality of life in sickle cell disease. *Journal of Clinical Outcomes Management, 22*, 10–20.

Treadwell, M. J., Barreda, F., Major, K., Walker, V., Payton, W., Kaur, K., et al. (2011). Mental health symptoms, quality of life and barriers to accessing health care in sickle cell disease. *Blood, 118*, 337. https://doi.org/10.1182/blood.V118.21.337.337, 337.

Treadwell, M. J., Hassell, K., Levine, R., & Keller, S. (2014). Adult sickle cell quality-of-life measurement information system (ASCQ-Me): Conceptual model based on review of the literature and formative research. *The Clinical Journal of Pain, 30*, 902–914. https://doi.org/10.1097/AJP.0000000000000054

Treadwell, M. J., Mushiana, S., Badawy, S. M., Preiss, L., King, A. A., Kroner, B., et al. (2022). An evaluation of patient-reported outcomes in sickle cell disease within a conceptual model. *Quality of Life Research, 31*, 2681–2694. https://doi.org/10.1007/s11136-022-03132-z

Tsyvkin, E., Riessman, C., & Mathew, P. (2015). Distrust and conflict in sickle cell disease: Intersecting narratives of patients and physicians. *Blood, 126*, 4472. https://doi.org/10.1182/blood.V126.23.4472.4472, 4472.

Wallen, G. R., Minniti, C. P., Krumlauf, M., Eckes, E., Allen, D., Oguhebe, A., et al. (2014). Sleep disturbance, depression and pain in adults with sickle cell disease. *BMC Psychiatry, 14*, 207. https://doi.org/10.1186/1471-244X-14-207

Weatherall, A., Keevallik, L., La, J., Dowell, T., & Stubbe, M. (2021). The multimodality and temporality of pain displays. *Language & Communication, 80*, 56–70. https://doi.org/10.1016/j.langcom.2021.05.008

Young, A. J., Richardson, F., Fitzgerald, D., Heavrin, B. S., Tweddell, B., Gettings, L., et al. (2020). Let their voices be seen. *Annals of Emergency Medicine, 76*, S73–S77. https://doi.org/10.1016/j.annemergmed.2020.08.015

Zaidi, A. U. (2021). "I can't breathe": How sickle cell disease manifests in the USA today. *Lancet Haematol, 8*, e479–e480. https://doi.org/10.1016/S2352-3026(21)00131-9