Sickle cell disease (SCD) is a chronic condition that affects thousands of people worldwide. The purpose of this study is to illustrate some of the challenges and perceptions of people diagnosed with SCD. The aims were to describe stressors, problematic symptoms, sources of support, and interactions with healthcare providers. This descriptive study, using mostly open-ended questions and conceptual analysis, included a sample of patients with SCD who were older than 18 years in ambulatory (57%) and inpatient (43%) clinics. Participants completed a 20-minute interview. Pain or SCD crisis were the chief reasons for hospitalization, and a wellness checkup was the chief reason people returned to an ambulatory clinic. Most (74%) were able to perform chores. Family was reported to cause the most stress in the home and also provide the most help. Pain is a pervasive aspect of life, limiting activities of daily living. Negative interactions with healthcare providers are common; therefore, advocacy is necessary for patients with SCD.
with sequelae of chronic pain and treatment. In addition to physical complications, about 30% of patients with SCD are diagnosed with depression (Sogutlu, Levenson, McClish, Rosef, & Smith, 2011), and many have other undiagnosed psychosocial concerns. Adults diagnosed with SCD screen for a higher rate of depression as compared to the overall U.S. population (Jenerette, Funk, & Murdaugh, 2005). In addition, people living with SCD tend to have moderate rates of reported suicidal ideations and require care from mental health professionals (Edwards et al., 2009).

**Frequent Hospitalizations**

Frequent and prolonged hospitalizations (Shankar et al., 2005), a history of severe pain crisis, regular emergency department (ED) visits, and related complications of SCD are associated with an increased probability of mortality (Houston-Yu, Rana, Beyer, & Castro, 2003). Hospitalization and readmission are both high, with the most substantial rates occurring among those ages 18–30 years old. Young adults are particularly at risk for complications and multiple hospitalizations as they transition from pediatric to adult care (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010). Among 108 people diagnosed with SCD, 78 were hospitalized within a four-year period of time with a mean hospitalization rate of five days; however, some were admitted as many as 20 times (Houston-Yu et al., 2003).

Sometimes people say, “You’re at the hospital again? . . . You just got out!” People don’t seem to understand. It’s not some sort of a vacation or something I enjoy doing. The pain feels like a bad toothache in every bone in my body. Why else would I want to come to the hospital? I do everything I can to keep from coming in: warm baths, distraction, compresses, resting, fluids, and home meds. Eventually nothing helps, and I have little choice. I hate being away from home and my little girl. Sometimes I don’t have anyone to take her, so I spend much of the hospitalization worrying and trying to get a sitter. I gather my strength and ask to be discharged, knowing my daughter needs me and knowing I’ll need readmitted soon.

**Attitudes of Healthcare Providers**

Patients who present to the ED for pain associated with SCD have an extended wait time of 25% longer than the general patient population (Haywood, Tanabe, Naik, Beach, & Lanzkron, 2013). Some of the wait time may be associated with the African American race (Haywood et al., 2013). Disparities in care and ED wait times cause patients with SCD to experience significant delays in treatment and analgesia care (Tanabe et al., 2007). Racial prejudices are not uncommon to people who are diagnosed with SCD and require frequent care in EDs and hospitals (Haywood et al., 2013; Weisberg, Balf-Soran, Becker, Brown, & Sledge, 2013). Patients, families, and healthcare providers feel racism affects the quality of health care provided in the ED (Nelson & Hackman, 2013). Perceptions that SCD is often ignored or unvalued are not uncommon (Aljuburi et al., 2012).

ED providers who admit to harboring negative attitudes toward patients with SCD are more likely to undertreat complaints of pain (Glassberg, Tanabe, Chow, et al., 2013). Many providers report feeling as though patients exaggerate discomfort, fail to comply with medical advice, abuse drugs, or are manipulative (Ratanawongsa et al., 2009); however, many patients with SCD wait until pain is reported at 8 on a 10-point scale before seeking care (Jenerette, Brewer, & Ataga, 2013). Patients who tend to have more hospitalizations and encounter staff disputes often inspire negative attitudes in their healthcare providers (Ratanawongsa et al., 2009). Many patients with SCD loathe being admitted to a hospital, and avoiding hospitalization is a major theme associated with patients seeking care for pain control (Jenerette et al., 2013).

The manner in which some healthcare providers refer to people diagnosed with SCD as “sicklers” can be reflective of negative attitudes and a lack of adherence to clinical guidelines associated with the disease (Glassberg, Tanabe, Richardson, & DeBaun, 2013). Patients with SCD commonly believe that providers doubt their reports of pain or perceive that providers do not have much experience with treating SCD (Jenerette, Leak, & Sandelowski, 2011).

Is it the pain or the color of my skin? What is it that makes some people want so badly to help, and others [to] just call me a drug seeker or worse . . . an addict? Others try to say things like, “I’m afraid to give you that much medicine.” Have they looked to see how much I have gotten? Is it an excuse? What can I say to make them understand? What about the awful pain? What do I do about that? What can I say? Who will listen?

**Methods**

This descriptive study used open-ended questions and a conceptual analysis. The specific aims were (a) to describe perceptions of stressors, problematic symptoms, and sources of support in people who are diagnosed with SCD and (b) to describe patients’ perceptions of experiences and interactions with healthcare providers.

Participants were diagnosed with SCD and receiving care in either an inpatient or ambulatory care setting. All people diagnosed with SCD older than 18 years of age were invited to participate. Patients were excluded from participation if they were younger than 18 years of age, unable to understand the consent forms, or incapable of reading or speaking English. The study was conducted at the Arthur G. James Cancer Hospital and Richard J. Solove Research Institute of the Ohio State University Comprehensive Cancer Center in Columbus. Approval was obtained from the institutional review board of the university.

**Procedures**

Patients were approached and invited to participate in the study while awaiting a clinic visit or in the hospital. Informed consent was obtained prior to participation. Demographic items, such as age, complications, and number of reported hospital admissions during the past year, were collected.

The investigator team included a clinical nurse specialist (CNS), direct care nurses, a psychiatric mental health CNS, and a nurse researcher. A trained research assistant (RA) conducted a 20-minute interview that consisted of five open-ended items.
and one closed-ended item (see Figure 1). Patient responses were written by the RA on a hard copy form. If patients requested, they were provided with writing materials to record their stories.

Analysis

Nine participants wrote their own narratives, and 68 of the interviews were recorded by the RA. The interview data were a paragraph-length response to the six items that guided the patient interviews. Each member of the investigator team reviewed all of the interview data, met and discussed the data, and reached consensus on the themes and the use of specific interview excerpts used in the results. The existence of themes was established from the data by recognizing similar words or phrases. All identified themes were agreed on by the team. Responses were grouped according to the guiding theme. A content analysis was conducted on the survey responses, and frequencies of themes were recorded (Laurence & Margolis, 2003).

Results

Of the 77 participants, most were women, and the mean age was 30 years (see Table 1). The mean number of reported complications was 1.97 (see Table 2). In addition, 57 participants said they were able to perform their daily activities without assistance (see Table 3). In regard to hospitalization, 21 participants attributed it to pain, whereas 14 participants said it was caused by SCD crisis. The nature of stress in the home was most often reported to be family. Likewise, the most frequently reported source of support or help in the home was family or friends. Reasons for frequent hospitalizations or clinic visits were pain, SCD crisis, medications, symptoms such as weakness and dehydration, and sometimes simply wellness checkups.

I came to the hospital with SCD pain because I overexerted myself with rigorous dance steps.

Pain in SCD often comes on suddenly and crescendos into a crippling and consuming problem that sometimes warrants hospitalization. Pain can escalate to the extent that many patients fail to get relief from oral analgesia. Although pain was described differently, the theme of severity, the need for medical attention, and the extent to which life is controlled by the pain were issues that patients frequently mentioned.

I can be having a good day and, all of a sudden, my pain [appears].

I have a bad hip. It triggers a crisis: lower back pain and tailbone spasms to [my] legs.

I work three jobs, so I overdo it. I put in my two weeks’ notice for one of my jobs.

Pain was reported many times as controlling various facets of life. People who are diagnosed with SCD report having to contend with pain even while carrying out their daily tasks and responsibilities.

At home, the pain level was up to 5, which is normal. It is never zero. I cope, take care of the home [and] kids, and take the few medications at home. Activities keep us occupied from the pain. When [pain] is over 5, it’s harder to take care of the kids. Three out of seven days [a week] will be normal days, and the rest will be hard.

Interaction with healthcare providers can be a struggle for many people diagnosed with SCD. Patients who require emergency care and frequent hospitalizations report feeling that providers do not believe they are in pain or the severity of their pain, or think they are labeled for addiction issues. Pain is a divisive symptom between patients and the healthcare team.

The doctors and nurses need to have more considerations to the patients’ pain. Some don’t believe in the pain. We’ve [the patient and her sister] had the disease all our life. We know from the type of pain if it’s due to anemia, dehydration, or pain crisis. We need doctors and nurses to be partners with us.

Many times I get comments from nurses and medics that I’m coming in to the hospital for medication for an addiction as their first thought. It would improve care if doctors didn’t have that in their brain.

Often, stressors associated with health or family motivate a crisis, and patients can tell when they are about to occur.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>X</th>
<th>SD</th>
<th>Range</th>
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<tbody>
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<td>19–59</td>
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<table>
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</tr>
<tr>
<td>Male</td>
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<table>
<thead>
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<tbody>
<tr>
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<tr>
<td>Inpatient</td>
<td>33</td>
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<table>
<thead>
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<th>Number of hospital visits</th>
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<td>37</td>
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<td>1</td>
<td>13</td>
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<td>2–3</td>
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<td>16–30</td>
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</tr>
<tr>
<td>50</td>
<td>1</td>
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</table>

TABLE 1. Sample Characteristics (N = 77)
I was discharged from the hospital after pneumonia, and the doctor thought it may cause a crisis. Soon after, I went [back to] the hospital, and I went into crisis.

Stressors that are common for people diagnosed with SCD are family concerns, financial issues, the frustration of being a caregiver to others while not feeling well, and worries about maintaining employment. For some, the loss of others to complications associated with SCD is also a reality.

My identical twin sister passed away from SCD, and I have her children, too.

Right now, I’m not working. I’m worried about the bills and my health. I am also worried about my daughter; she has cerebral palsy.

I have a baby at home. [I] just broke up with [the] child’s father, and now I’m financially responsible for the apartment.

[My] brother passed away. [It] feels like no one in my family loves me.

Families often provide support to people diagnosed with SCD. Although mothers, fathers, siblings, grandparents, friends, and significant others all were named by participants as sources of support, many patients did not have anyone to assist with their care and daily activities. For some, families were available but not supportive.

Honestly, my family and friends have become so immune to this lifelong struggle that they don’t really get involved with assisting me. On the other hand, I live two hours away from them, too, so by the time I inform them of a possible need for help, it is too late. And I, like most people I know with [SCD], have become severely independent and handle my health needs alone.

### Discussion

SCD is a condition that people must endure for their entire lives. In addition to the disease itself, patients with SCD have to cope with many difficult issues, such as the deaths of other family members, financial problems, and their invisibility to the healthcare system. Patients with SCD have families, go to school and work, have friends, and function in society. Many studies have also illustrated some of the stressors and physical challenges associated with SCD in adults, children, and families (Dos Santos & Gomes, 2013; Hildenbrand, Barakat, Alderfer, & Marsac, 2013). Complications associated with SCD are rather common and increase in incidence with aging (Blinder et al., 2013). Despite the relatively young mean age of the sample, the average number of complications associated with SCD was two, and 37 participants did not require a hospitalization in the past year. This is high, as a large multistate study found that about 30% of patients with SCD did not require hospitalization in the span of a year (Brousseau et al., 2010). When patients with SCD present to the ED, about half require hospitalization (Po’ et al., 2013).

Pain is a central theme in many of the participant responses and in much of the published research about SCD (Jenerette et al., 2013; Platt et al., 1991; Solomon, 2010). The unpredictability and life-altering nature of how pain is encountered and endured was discussed frequently (Pereira, Brener, Cardoso, & Proietti, 2013). Pain was often referred to as a common aspect of life and depicted as a barrier to living a normal life.

Participants were aware that many providers did not believe their reports of pain. Unless distress was visible, participants felt they were not going to be believed in regard to the extent of pain. This issue is also represented in the literature as a particular problem for patients with SCD and as an area of education for healthcare providers (Haywood, 2013).

Presumptions of drug seeking and addiction were pervasive among healthcare providers and palpable to the participants (Haywood, 2013; Jenerette et al., 2013). The research team believed that the issue of race was an unspoken theme in the interviews. Racial or socioeconomic prejudices that have been documented in the literature were never mentioned. Perhaps because the team consisted of Caucasian women, many patients did not feel comfortable discussing feelings of race.

Many stressors were described as inevitable in living with SCD. Deaths of relatives and friends that resulted from SCD were often described. Problems with family members and significant others were common, as were financial problems because of difficulty maintaining employment. Family members were reported as providing the most help and yet the largest source of stress. Caring for others while not feeling well is another aspect of life with SCD. A significant psychological burden does exist for people diagnosed with SCD (Jonassaint, Jonassaint, Stanton, De Castro, & Royal, 2010).

### TABLE 2. Complications of Sickle Cell Disease (N = 77)

<table>
<thead>
<tr>
<th>Complication</th>
<th>n</th>
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<tbody>
<tr>
<td>Acute chest syndrome</td>
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<tr>
<td>Cardiac complications</td>
<td></td>
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<td>8</td>
</tr>
<tr>
<td>No</td>
<td>48</td>
</tr>
<tr>
<td>Missing</td>
<td>21</td>
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<tr>
<td>Chronic pain medicine</td>
<td></td>
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<tr>
<td>Yes</td>
<td>57</td>
</tr>
<tr>
<td>No</td>
<td>20</td>
</tr>
<tr>
<td>Iron overload</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>24</td>
</tr>
<tr>
<td>No</td>
<td>47</td>
</tr>
<tr>
<td>Missing</td>
<td>6</td>
</tr>
<tr>
<td>Liver insufficiency</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>4</td>
</tr>
<tr>
<td>No</td>
<td>67</td>
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<tr>
<td>Missing</td>
<td>6</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>12</td>
</tr>
<tr>
<td>No</td>
<td>59</td>
</tr>
<tr>
<td>Missing</td>
<td>6</td>
</tr>
<tr>
<td>Renal insufficiency</td>
<td></td>
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<tr>
<td>Yes</td>
<td>8</td>
</tr>
<tr>
<td>No</td>
<td>61</td>
</tr>
<tr>
<td>Missing</td>
<td>8</td>
</tr>
</tbody>
</table>
A limitation of the study was that the authors did not collect data on the type of SCD with which participants had been diagnosed. Common types of SCD include HbSS, or sickle cell anemia, which is typically the most severe form of the disease and is caused by the inheritance of one sickle cell gene from each parent, and HBSC, which is usually a milder form of SCD and results from the inheritance of a sickle cell gene from one parent and a gene for an abnormal hemoglobin from the other.

In addition, in regard to the study, the authors failed to audio record the interviews. A consultant to the project suggested that many people would be averse to having their communications recorded and, therefore, not give a true story or an entire story. The participants in this study were very willing to talk and likely would have had no problem with recording the interviews. Participants (n = 9) wrote their stories on the backs of the hard copy forms and continued to motivate discussions beyond the structured interviews.

Implications for Nursing Practice

Nurses must be the advocates for patients who require care for SCD. They must understand that pain, frequent ED visits, and hospitalizations are often part of the disease, and that they should not inspire negative perceptions and poor-quality health care. Nurses must note these insufficiencies (Haywood, 2013) and help to cultivate a more positive climate in the healthcare setting for people diagnosed with SCD. Resources are available to help clinicians improve their attitudes toward caring for patients with SCD.

Pain control is a major aspect of care for patients with SCD. Nurses, who are able to translate nonpharmacologic pain and symptom management techniques in the care of patients who are diagnosed with SCD, must advocate for better pain control and shorter wait times for pain medications in the ED. They must also ameliorate other elements of well-documented disparities.

Conclusions

Pain or an SCD crisis were the most common causes of hospitalization or clinical visits. Pain is a pervasive aspect of life for many people with SCD. Negative interactions with healthcare providers are common, particularly if the patient requires analgesia. Family concerns, financial issues, and patient caregiving responsibilities are some of the stressors frequently encountered, as are concerns over maintaining employment or staying in school. Although family may sometimes be a source of stress, family is also reported to be the largest provider of support.

Sensitivity and empathy are elements of nursing that are frequently employed for patients and families with chronic conditions, a diagnosis of cancer, or end-of-life care (Haywood, 2013). The same care and concern must be extended to all patients, no matter what disease they have or from what ethnic background they come. Supporting the family and caregivers of patients with SCD is a critical role of the nurse.

References


<table>
<thead>
<tr>
<th>TABLE 3. Participant Responses Regarding Visit (N = 77)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Question and Response</td>
</tr>
<tr>
<td>What brings you to the hospital or clinic?</td>
</tr>
<tr>
<td>Checkup</td>
</tr>
<tr>
<td>Pain</td>
</tr>
<tr>
<td>Sickle cell disease crisis</td>
</tr>
<tr>
<td>Anemia</td>
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<tr>
<td>Dehydration</td>
</tr>
<tr>
<td>Weakness</td>
</tr>
<tr>
<td>Pain medications</td>
</tr>
<tr>
<td>Other</td>
</tr>
<tr>
<td>What is your ability to perform chores?</td>
</tr>
<tr>
<td>Able</td>
</tr>
<tr>
<td>Able with assistance</td>
</tr>
<tr>
<td>Unable</td>
</tr>
<tr>
<td>What causes stress in your home?</td>
</tr>
<tr>
<td>Family</td>
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<td>Other</td>
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<td>Chores</td>
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<td>Pain or sickness</td>
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<td>Hospital visits</td>
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<td>No support</td>
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<td>Staff</td>
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Note. Participants could select as many or as few responses as they wished, so totals do not equal N.


