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Sickle Cell Disease in Adults: Developing an Appropriate Care Plan

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Abstract

Background—Sickle cell disease (SCD) is primarily characterized by pain. This chronic pain with acute exacerbations is the most common reason for hospital visits, admissions, and readmissions, particularly in young adults (aged 18–39 years). People who present to the hospital for pain crises often report that nurses lack knowledge of SCD and, consequently, they do not provide appropriate, timely care.

Objectives—Because pain episodes often result in hospital admissions, this article highlights prominent issues that staff nurses need to know.

Methods—Using a review of the literature and case studies, the authors provide recommendations to improve care of adults with SCD.

Findings—No objective signs of a sickle cell pain crisis exist. Patients react to pain in different ways and use various coping mechanisms in response. Suspected opioid addiction should not affect the provision of nursing care. Pain must be treated appropriately to decrease the potential for prolonged admissions and/or readmissions. Patients are to be acknowledged as experts and collaborated with in developing an appropriate plan of care. Advocacy on behalf of the patient is important for better communication with providers. With this knowledge, nurses will be better equipped to provide the appropriate and timely care required to manage pain crises experienced by individuals living with SCD.

Graphical Abstract

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Keywords

sickle cell disease; sickle cell anemia; sickle cell pain; clinical management; pain; staff nurses

Adults with sickle cell disease (SCD) average more than 197,000 annual visits to the emergency department (ED), with 29% of visits resulting in hospital admissions (Yusuf, Atrash, Grosse, Parker, & Grant, 2010). These visits often are less than ideal. Patients with SCD and their families report feeling that race affects healthcare quality and interpersonal relationships with caregivers, and results in negatively different treatment (Nelson & Hackman, 2013). When they sought pain treatment at hospitals, they reported being stigmatized as drug seekers (Jenerette, Brewer, & Ataga, 2014) and that their experiences often involved mistrust by providers, lack of patient control, and neglect (Prabhakar, Haywood, & Molokie, 2010). Patients with SCD also encountered underestimation, misunderstanding, and inadequate management of their pain (Zempsky, 2010). They experienced long delays in receiving pain medications and providers who lacked an understanding of SCD (Lattimer et al., 2010). Lattimer et al. (2010) also reported that patients with SCD felt that they were not always treated with respect and dignity, they were inadequately involved in decisions about their care, they received conflicting information from staff, nurses did not provide clear answers to their questions, finding someone with whom to discuss concerns was difficult, and their fears and anxieties were not always discussed by the nurses. In addition, some patients reported that their family was not given enough information to help with their recovery (Lattimer et al., 2010).

Every staff nurse must know certain things about SCD and integrate them into practice to provide appropriate and timely care for individuals living with SCD. The purpose of this article is to help staff nurses better understand SCD and to highlight prominent issues associated with SCD that will improve clinical practice, resulting in improved health outcomes for individuals living with SCD. This information is particularly important for oncology nurses who often care for individuals with SCD, a hematologic disorder, because they have expertise in pain assessment and management and understand that SCD symptomatology and therapies are similar to those commonly seen in some cancers (Jenerette & Leak, 2012).

Understanding Sickle Cell Disease

SCD is an autosomal recessive disorder caused by an amino acid substitution (from glutamine to valine) on the beta globin gene (Brousse, Makani, & Rees, 2014). This point mutation results in a distortion in the hemoglobin (Hgb) structure upon deoxygenation, causing the red blood cells (RBCs) to become sickle-shaped. These sickle-shaped cells adhere to each other and to the blood vessel walls, resulting in blocked blood flow and decreased circulation to body tissues, irreversible organ damage, and various complications such as inflammation and pain (Olowoyeye & Okwundu, 2014). The three most common types of SCD include HbSS (known as sickle cell anemia), HbSC, and HbS beta (0 and +) thalassemia, with HbSS being the most severe (aCenters for Disease Control and Prevention [CDC], 2014a). Compared to the average Hgb value of 13.8–17.2 g/dl for males and 12.1–15.1 g/dl for females (U.S. National Library of Medicine, 2015), an individual with SCD has an average Hgb value of 6–9 g/dl (Howard & Oteng-Ntim, 2012; MacMullen & Dulski, 2011).

Individuals of African, Caribbean, Central and South American, Saudi Arabian, Indian, and Mediterranean descent experience the highest rates of SCD (CDC, 2011). In the United States, about 100,000 individuals are affected (CDC, 2011), making it the most common genetic blood disorder (Genetics Home Reference, 2012). The majority of these individuals are African American. They experience a disease incidence of 1 in 500 births and the presence of the trait in 1 in 12 births (CDC, 2011). Males and females are affected equally (Pack-Mabien & Haynes, 2009). Testing for SCD can be done after the second month of a pregnancy (CDC, 2014c) and is included in newborn screenings (CDC, 2014b).

The only cure for SCD is bone marrow or stem cell transplantation (CDC, 2014a). Unfortunately, this is not a viable treatment option for many because few potential donors are available (Ataga, 2009). This procedure is optimal during childhood and in those with less advanced cases, but it can have fatal side effects (Buchanan, Vichinsky, Krishnamurti, & Shenoy, 2010).

Complications

Many symptoms and disease complications are associated with SCD that range in severity and vary by individual. The most common symptom is chronic and/or acute pain (Ballas et al., 2012). Pain is associated with disease complications (often labeled “pain syndromes”) and in itself is a major complication of SCD. Pain crises (also known as pain episodes, sickle cell crises, or vaso-occlusive crises) are periods of acute, severe, bony pain that occur throughout life and are unpredictable (Howard & Oteng-Ntim, 2012). These crises are attributed to many causes that differ for each person and may primarily include sickling of the RBCs, vaso-occlusion, ischemia, infarction, inflammation, and tissue damage (Ballas et al., 2012; Darbari, Ballas, & Clauw, 2014). Individuals with SCD often associate stress, weather changes, cold exposure, dehydration, fatigue, and overexertion with pain onset, but most pain crises are not associated with a clear precipitating factor (Ballas et al., 2012). Pain locations vary, but the extremities and the back are most affected (Brousse et al., 2014). On average, individuals experience at least one pain crisis annually (Taylor, Stotts, Humphreys,

Treadwell, & Miaskowski, 2010); however, they can occur as frequently as weekly or monthly (Ballas et al., 2012). A typical crisis lasts about 10 days, with women reporting longer episodes than men. They are most frequent in people aged 19–39 years, are the most common reason for hospital visits (Yusuf et al., 2010), and cause 90% of SCD-related hospitalizations (Dunlop & Bennett, 2006). The frequency of pain crises is associated with premature death in people with SCD aged older than 20 years and, therefore, is indicative of disease severity (Pack-Mabien & Haynes, 2009). In addition, the mortality risk in this population increases with the number and duration of hospitalization days per year (Houston-Yu, Rana, Beyer, & Castro, 2003).

Chronic pain occurs between pain crises and is usually considered milder (Ballas et al., 2012). Pain locations may include the head, neck, chest, back, abdomen, or limbs. The pain can last for months or years and is accompanied by suffering, anxiety, despair, helplessness, depression, insomnia, and loneliness. Other common complications that can be seen with a pain crisis include acute chest syndrome, pulmonary hypertension, sickle nephropathy, ischemic stroke, avascular necrosis, leg ulcers, infection, heart disease, and renal failure. Acute chest syndrome is the second most common reason for hospitalization in people with SCD and, along with pulmonary hypertension, causes the highest mortality rates in this population (Miller & Gladwin, 2012).

Management

Management of SCD involves focusing on the treatment of pain and preventing complications (Lee, Askew, Walker, Stephen, & Robertson-Artwork, 2012). A typical pain medication regimen includes opioid analgesics (e.g., morphine, hydromorphone [Dilaudid®], oxycodone [OxyContin®]) and nonopioid analgesics (e.g., ibuprofen, acetaminophen) (Niscola, Sorrentino, Scaramucci, Fabritiis, & Cianciulli, 2009; Telfer, Bahal, Lo, & Challands, 2014). In addition to pain medications, RBC transfusions and hydroxyurea (Hydrea®) may be used to help reduce levels of Hgb S (Dunlop & Bennett, 2006; Pack-Mabien & Haynes, 2009). These therapies help decrease pain, but they do not completely prevent crises. Management of SCD occurs primarily at home and depends on self-care. Self-care strategies include the aforementioned pharmacologic methods, as well as nonpharmacologic methods (e.g., warmth, hydration, rest, good food, avoiding alcohol and drugs, listening to and learning about the body, prayer, social support) (Jenerette, Brewer, & Leak, 2011).

When a pain crisis arises and self-care strategies are no longer effective in achieving pain relief at home, individuals seek care at the hospital (Jenerette, Brewer, & Ataga, 2014). There, they typically receive IV fluids, oxygen, and pain medications and then are discharged when they are considered stabilized; however, the crisis may still be ongoing (Ballas, Gupta, & Adams-Graves, 2012). More severe cases or those that have progressed because of delay in the administration of analgesia may result in hospital admission for additional symptom management and/or treatment of the underlying problem (Ballas, 2011). See Figure 1 for additional evidence-based evaluation and management guidelines.

Challenges in Managing Pain

No objective signs of a sickle cell pain crisis—A 19-year-old male with SCD said,

He checked my blood work and then came back to me and told me that I wasn't in a pain crisis. They said that my hemoglobin count was higher than what a person in a crisis would be. Mind you, my hemoglobin count runs 15 and, like, the average person with sickle cell runs at 10, so I'm already at a high blood count.

The pain crisis experience is unique to each individual. Individuals often present with severe pain, usually a pain rating of more than 6 on a scale of 0–10 (Ballas et al., 2012). The pain is typically described as throbbing, sharp, dull, or stabbing and is most often located in the back, legs, knees, arms, chest, and abdomen. Objective findings supporting these pain reports and the presence of a pain crisis often do not exist. In addition, individuals may not present as expected. For example, one might expect the Hgb level to be inversely related to pain. In actuality, individuals with sickle cell anemia who have relatively high Hgb levels are prone to more frequent pain crises than those with lower Hgb levels (Ballas et al., 2012). Given the individual nature of disease symptomatology and the absence of objective findings, nurses must depend on the patient's report to effectively assess and manage pain.

Pain is a subjective experience that manifests in various ways. Nurses are taught that pain is the fifth vital sign and that pain is “whatever the experiencing person says it is, existing whenever the experiencing person says it does” (McCaffery, 1968, p. 95). In adults, a numeric rating scale (0–10) is most often used to assess pain. In patients with SCD, this subjective scale should be used to measure pain and set goals for pain management. Remember that pain ratings are not the same for each individual. For example, some patients may be ready for discharge at a pain rating of 5, while others may not. Patient-provider communication is key for mutual goal setting and achieving optimal pain outcomes.

A 32-year-old male with SCD said,

Sometimes me and the nurses clash. I know how my medicine should be given. ... Some of them don't really understand the pain that sickle cell patients are in. So it's very stressful to even be in the hospital. And then I know that being stressed out will keep me in there longer.

Patients with sickle cell disease cope differently in response to pain—

According to a 36-year-old woman who has been a BSN-prepared nurse for six years,

Rating pain at an 8 out of 10 when the patient is laughing, talking on the phone, and socializing with friends does not warrant additional doses of narcotics. There needs to be a better gauge than what the patient reports pain to be.

Successfully living with lifelong pain requires the use of coping strategies. These strategies may be active (e.g., increasing activity) or passive (e.g., hoping the pain goes away) (Higgins, Bailey, LaChapelle, Harman, & Hadjistavropoulos, 2015). In addition, some people may cope more effectively than others. For instance, pain may be viewed as fixed or malleable. Individuals who believe that pain is fixed tend to use fewer active coping strategies, express greater pain levels, and show more facial signs of pain. Social support

from family and friends is also important because it is associated with higher levels of resilience (Newton-John, Mason, & Hunter, 2014) and, therefore, more adaptive coping (Jenerette, 2008).

Pain expressions may not always reflect pain presence and intensity because individuals perceive, describe, and deal with pain differently (Shavers, Bakos, & Sheppard, 2010). Patients in pain may respond by crying, grimacing, laughing, or by simply ignoring the pain. Individuals with SCD tend to use more passive coping, such as prayer (Thompson & Eriator, 2014) and spirituality (Taylor, Stotts, Humphreys, Treadwell, & Miaskowski, 2013), in response to pain. Young adults, in particular, tend to use a mixture of active and passive strategies. They often ignore the pain or use heat, cold, and massage to cope (Sanders, Labott, Molokie, Shelby, & Desimone, 2010). Distraction (e.g., listening to music, talking on the phone, talking with visitors) also is used to cope. Nurses cannot rely on their expectations of what pain looks like or should look like in determining pain presence because doing so is problematic to assessing pain. The nurse must listen to individuals with SCD, believe their pain reports, and encourage the use of positive coping strategies as supplements to pharmacologic management.

Tolerance, physical dependence, pseudo-addiction, or addiction?—A 19-year-old female with SCD said,

They will label you as addicted or whatever to the pain medication. But that's not the case because you are in pain and you just want the pain to go away. ... It's just difficult to get the help that you need.

High doses of potent analgesics are often necessary to address the pain experienced by individuals living with SCD. When they access the healthcare system, these individuals may request specific medications that have been successful in managing their pain in the past. Nurses should expect specific requests because the patients are the content experts, are informed consumers, and are involved in self-care for pain management. This behavior, combined with the absence of objective pain signs, is often incorrectly perceived as drug seeking, opioid dependency, or addiction (Nelson & Hackman, 2013; Shavers et al., 2010). The nurse's lack of knowledge may lead to dependence on stereotypes, personal beliefs, and attitudes, which often do not accurately reflect the patient or his or her behavior; therefore, patient care is negatively affected (Shavers et al., 2010). In addition, empathy biases have been noted in health care. These biases refer to favoritism shown by providers toward individuals with whom they identify, often based on race, ethnicity, or gender (Drwecki, Moore, Ward, & Prkachin, 2011).

Case Study

Marcus, a 21-year-old male with sickle cell disease (SCD), has been experiencing pain in his back and legs at home for the past three days. He has been resting, using a heating pad, taking his prescription pain medications, and drinking lots of water in an attempt to address the pain. These strategies are not working, and the pain has become severe—a 10 on a scale of 0–10. He no longer can control the pain at home, so he decides to go to the hospital.

His persistent pain and hemodynamic instability result in admission to an inpatient unit for additional evaluation and management. As a staff nurse on this unit, you are assigned to care for Marcus. His admitting diagnoses are sickle cell anemia and pain crisis. What do you know about SCD? What will his plan of care entail?

Developing a Care Plan to Manage Pain

Let's consider the appropriate plan of care for Marcus. Once he's admitted to your unit, Marcus should be fully assessed for pain. Determine the location, intensity, characteristics, and associated symptoms (National Heart, Lung, and Blood Institute, 2014). Afterward, collaborate with Marcus to identify pain management strategies (pharmacologic and nonpharmacologic) that were most effective in the past. Identify and initiate the best strategies for the current situation.

Pain management (with opioids and nonsteroidal anti-inflammatory drugs), hydration, and oxygenation are primary goals of care (Gregory, 2012). Once pain management is initiated via patient-controlled analgesia, reassess Marcus's pain every 15–30 minutes until he reports that the pain is under control (National Heart, Lung, and Blood Institute, 2014). Encourage the use of distraction, local heat, incentive spirometry, and ambulation and activity as soon as possible.

While employing these strategies, identify and directly address the underlying cause of pain. Once Marcus is hemodynamically stable and has reached a pain level that he finds tolerable and manageable at home, begin discharge. Wean him off the parenteral medications in preparation for conversion to oral medications, discuss use of self-care strategies to manage pain at home, and provide him with resources for outpatient follow-up if needed (National Heart, Lung, and Blood Institute, 2014).

An unfounded and unwarranted concern exists among nurses regarding addiction in people with SCD. Nurses believe that most patients with SCD are drug addicts (Pack-Mabien, Labbe, Herbert, & Haynes, 2001). In fact, addiction risk in this population is low at less than 10% (Solomon, 2008). Nurses must understand the differences among tolerance, physical dependence, pseudo-addiction, and addiction to treat individuals living with SCD appropriately. With repeated doses of the same analgesics, as can be expected in cases of lifelong pain, individuals can become tolerant or develop a decreased response over time, seen as a shortened duration of analgesic effectiveness (Smith, Jordan, & Hassell, 2011). Higher doses of analgesics then are required to achieve relief. Physical dependence refers to the occurrence of withdrawal when an opioid agonist is stopped abruptly or when an antagonist is administered. Tolerance and physical dependence are as likely to be seen in individuals living with SCD as in anyone with a history of chronic pain and long-term opioid use (Solomon, 2008). Nonetheless, this possibility should not impair use of proper pharmacologic management. Pseudo-addiction refers to the seeking out of needed opioids, sometimes from multiple sources, along with increased pain behavior, anger, and acting out, resulting from uncontrolled pain and inadequate pain treatment (Weissman & Haddox, 1989). This can be seen in individuals with SCD and is often mistaken for addiction (Smith et al., 2011). One 35-year-old female with SCD said, "You have to act up to make people help you, and that ain't good."

Addiction refers to psychological dependence on opioids. This may be evidenced by use of opioids for reasons other than pain relief or the need for an increased dosage without increased pain (Smith et al., 2011). Suspected opioid addiction should not affect the provision of nursing care. Rather than rushing to judgment, nurses must work with patients to understand the reason for their behaviors and address them properly.

Nurses also should note that frequent visits to the ED with pain complaints and requests for potent analgesics do not necessarily constitute suspicious behavior. Rather, because of health challenges associated with SCD, individuals often are unable to work or maintain employment. They are more likely to be less educated and are more likely to be unemployed or disabled than African Americans without SCD (Laurence, George, & Woods, 2006). Financial limitations may arise because of a lack of education and employment or appropriate employment because of a lack of educational preparation (Matthie, Jenerette, & McMillan, 2015); therefore, the ED may be the primary source of care.

Implications for Nursing Practice and Education

Often described as the “iceberg phenomenon,” most of SCD pain is “submerged” at home and away from the healthcare setting, while only a small fraction of the pain, or the “tip of the iceberg,” is actually communicated to providers (Smith & Scherer, 2010). This may contribute to underestimation of pain by providers and result in ineffective treatment. When pain is treated inappropriately, the potential for prolonged admissions and/or readmissions increases. Guidelines exist for the care of patients with SCD experiencing pain crises. A nurse who follows these guidelines effectively manages pain and connects with the patients, their family members, and their caregivers. For more SCD resources, see Figure 2.

As direct providers of clinical care, nurses are in the best position to serve as patient advocates. To advocate for a patient, nurses must be aware of the patient’s needs and concerns for a true partnership dynamic with open communication. In addition, a good nurse-patient relationship is necessary for the nurse to empower the patient to be an active partner in his or her care (Doss, DePascal, & Hadley, 2011). Patients and their caregivers are to be acknowledged as experts, respected, and treated as valuable members of the healthcare team. Accordingly, care processes should include shared decision making among patients, their caregivers, and nurses. Nurses also should educate patients and their caregivers about the importance of quickly reporting problems, such as pain, so that they can be addressed before they become too severe.

The nurse’s role in the care of people with SCD is multidimensional. Nurses can best evaluate their needs, monitor for complications, help them improve their self-care skills and quality of life, advocate for better communication with providers, and advocate for implementation of individualized care plans (Agency for Healthcare Research and Quality, 2013). This will improve care quality and patient satisfaction with care, and may ultimately contribute to decreased hospital admissions and readmissions.

Conclusion

To provide the best care for adults living with SCD, nurses must put aside their personal biases, be educated about SCD, and use this knowledge to treat patients appropriately. In addition, educated nurses can advocate on patients' behalf, particularly when patients cannot advocate for themselves.

Oncology nurses can play a significant role on behalf of patients with SCD because they are experts in pain assessment and management (Jenerette & Leak, 2012). When their experience and knowledge are appropriately applied to the care of these individuals, outcomes—namely pain management—for this vulnerable population should improve.

Oncology nurses can be leaders or “champions” on behalf of patients with SCD. They can educate colleagues that no objective signs of a sickle cell pain crisis exist and that patients' pain reports should be believed. Oncology nurses value coping behaviors; therefore, they should not only respect that coping behaviors are individualized, but also should teach and optimize them. Just as with patients with cancer, suspected opioid addiction should not play a role in the provision of nursing care to individuals with SCD because pain must be treated appropriately to decrease the potential for prolonged admissions or readmissions and reduce patient suffering. Individuals living with chronic conditions like SCD should be acknowledged as experts in their care and collaborated with in developing appropriate care plans. Oncology nurses should lead by example by sharing their expertise with individuals living with SCD in a joint effort aimed at improving patient outcomes.

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Implications for Practice

- Obtain subjective reports of sickle cell disease (SCD) pain for appropriate pain management and evaluation of treatment.
- Develop care plans collaboratively with individuals with SCD and their family members.
- Advocate on behalf of the individual with SCD to improve the quality of care delivered.

- Believe the patient's pain report and optimize therapeutic outcomes to achieve adequate pain relief and improve the patient's quality of life.
- Evaluate for causes of pain other than a vaso-occlusive crisis while beginning to treat pain.
- Assess individuals with a hemoglobin concentration that is 2 g/dl or more below their baseline (or less than 6 g/dl when the baseline is unknown) for acute splenic sequestration, an aplastic episode, a delayed hemolytic transfusion reaction, acute chest syndrome, and infection.
- Choose an analgesic based on pain assessment, associated symptoms, outpatient analgesic use, patient knowledge of effective agents and doses, and past experience with side effects.
- Use an individualized prescribing and monitoring protocol (from the patient's sickle cell disease [SCD] provider) or an SCD-specific protocol whenever possible.
- In cases of severe pain, rapidly initiate treatment with parenteral opioids.
- Reassess pain and readminister opioids as needed for persistent severe pain every 15–30 minutes until pain is under control per patient report.
- Use patient-controlled analgesia or frequently scheduled doses rather than "as requested" administration.
- Monitor for excessive sedation.
- For individuals unable to drink fluids, provide IV hydration at a maintenance rate.
- Administer oxygen for individuals with an oxygen saturation level less than 95% on room air.
- Reduce the risk of acute chest syndrome by encouraging use of incentive spirometry while awake and encouraging ambulation and activity as soon as possible.
- Use supplemental nonpharmacologic approaches (e.g., local heat, distraction) to treat pain.
- Hospitalize individuals with a temperature of 103.1°F (39.5°C) or greater and those who appear ill for close observation and IV antibiotic therapy.
- In individuals with a fever that is accompanied by shortness of breath, tachypnea, cough, and/or rales, obtain an immediate chest x-ray to investigate for acute chest syndrome.
- If individuals have symptoms or signs suggestive of pulmonary hypertension, refer them for echocardiography.
- Individuals who have more than three sickle cell–associated moderate to severe pain crises in a 12-month period, sickle cell–associated pain that interferes with daily activities and quality of life, history of severe and/or recurrent acute chest syndrome, or severe symptomatic chronic anemia that interferes with daily activities or quality of life, should be treated with hydroxyurea therapy.
- Discontinue hydroxyurea therapy in females who are pregnant or breastfeeding.
- Blood transfusions are often used in cases of severe complications and prior to surgery.

FIGURE 1.

National Heart, Lung, and Blood Institute Guidelines for the Management of Sickle Cell Disease

Note. Based on information from National Heart, Lung, and Blood Institute, 2014.

American Sickle Cell Anemia Association<http://ascaa.org>**Centers for Disease Control and Prevention**

Sickle Cell Disease

<http://cdc.gov/ncbddd/sicklecell/index.html>**International Association of Sickle Cell Nurses and Physician Assistants**<http://iascnapa.org>**National Heart, Lung, and Blood Institute**

Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014

<http://nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines>

What Is Sickle Cell Disease?

<http://nhlbi.nih.gov/health/health-topics/topics/sca>**Sickle Cell Adult Provider Network**<http://scapn.net/ohana>**Sickle Cell Disease Association of America**<http://sicklecelldisease.org>**Sickle Cell Information Center**<https://scinfo.org>

FIGURE 2.
Sickle Cell Disease Resources