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# Barriers to Care for Persons with Sickle Cell Disease: The Case Manager's Opportunity to Improve Patient Outcomes

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## Abstract

**Purpose and Objectives—**The purpose of this discussion is to review the barriers to care for patients with sickle cell disease (SCD). Chronic pain and the perception of addiction, implicit bias, frequent hospitalizations and emergency department visits, clinician and patient knowledge deficit and sickle cell disease stigma all impede the ability to provide evidence-based care for patients with sickle cell disease. Case managers can coordinate and advocate for appropriate care that improves patient outcomes.

**Primary Practice Setting—**This discussion is relevant to case managers working with patients with SCD in the clinic, hospital and emergency department.

**Findings/Conclusions—**Case managers can serve an important advocacy role and intervene to improve the coordination of services and efficient use of resources. This will lead to improved quality of life and optimal healthcare utilization for persons with SCD.

**Implications for Case Management Practice—**As a constant member of the health care team, the case manager may be the only health care team member that has broad knowledge of the patient's experience of acute and chronic pain, usual state of health, social behavioral health needs, and how these factors may affect both in-patient and out-patient healthcare use and health outcomes. This paper explores the barriers to care and suggests specific interventions within the

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role of the case manager that can improve care delivered and ultimately contribute to improved patient outcomes. Specifically these interventions can improve communication among members of the health care team. Case manager interventions can guide coordination, prevent hospital re-admissions, reduce health care utilization and contribute to overall improved patient quality of life and health outcomes.

**Keywords**

sickle cell disease; case management; healthcare utilization

The case manager is in an excellent position to help coordinate the complex medical and social behavioral needs for patients with sickle cell disease (SCD) and advocate for improved evidenced based care directly impacting long term patient outcomes.

There are many barriers to care. These barriers include: clinician and patient knowledge deficit, chronic pain and the perception of addiction, frequent visits, SCD stigma, and implicit bias…. Individualized discharge plans with direct patient involvement can reduce barriers faced after discharge and reduce readmission rates.

The perception that patients with SCD have higher rates of opioid addiction is not uncommon among health care providers. There is no evidence to support the belief that patients with SCD are addicted to opiates, yet health care providers continually label SCD patients as addicted. These negative attitudes demonstrated by health care professionals interfere with proper pain management in SCD.

## Introduction

Sickle cell disease (SCD) is a severe inherited chronic medical condition that disproportionately affects primarily African American persons. About 90,000–100,000 Americans suffer from this severe condition, most of whom are low income and minorities (Hassell, 2010). Persons with SCD experience a multitude of medical complications and associated social behavioral health needs. The wide array of medical complications often requires coordination of services from many specialties including hematology, primary care, pulmonology, nephrology, orthopedics, pain management and psychiatry. Individuals with severe disease are often unable to work resulting in challenges with insurance, obtaining prescriptions and coordination of care (S. K. Smith, Johnston, Rutherford, Hollowell, & Tanabe, 2017). These complications significantly shorten their lifespan compared to the general population with age of death approximately 42 for males and 48 for females (Lanzkron, Carroll, & Haywood, 2013). Healthcare providers are often ill equipped to provide expert care for individuals living with SCD (National Heart, 2014). The case manager is in an excellent position to help coordinate the complex medical and social behavioral needs for patients with SCD and advocate for improved evidenced based care directly impacting long term patient outcomes.

## Case Study

James is a 20 year old black male recently admitted to the emergency department for sickle cell crisis. He is quiet and makes little eye contact and repeatedly asks for pain medication. He has recently moved, and has not re-established care with a sickle cell physician yet. The social worker in the emergency department conducted a social history and found that James had to drop out of college because of frequent hospitalizations and recently moved back home with his mom. He has been depressed and anxious and misses all his friends. He has not found work, does not have a car, lives in a rural area and is unaware of potential sources of disability. James receives minimal pain control in the hospital for his crisis and the case manager helps him to advocate for adequate pain relief. The case manager also helps James transition into caring for his own health since his mother had previously helped James navigate throughout the hospital. James is discharged four days later after the case manager develops a plan of care with the health care team.

The social worker makes an appointment with his primary care physician after discharge and refers him to his case manager in the community for follow up. The community case manager works with James and discusses options for returning to school, maintenance of health, applying for disability and learning about the local transportation options. Overall, James receives education, support and hope for living well with sickle cell disease.

Patients with SCD are admitted to the hospital at a much higher rate than persons with other chronic medical conditions (Jiang, Barrett, & Sheng, 2014). In 2012, people with SCD had the highest numbers of hospital stays with 4 or more stays in 12 months consisting of 57% of all hospital stays for Medicaid “super- utilizers” (Jiang et al., 2014). The most common admission diagnosis is vascular occlusive episodes (VOE) (National Heart, 2014). In addition to suffering from acute VOE, many patients with SCD also suffer from chronic daily pain limiting functional status, movement and activities and decreasing the quality of life. Treatment of VOE often requires high doses of opioids, leaving many health care providers (HCPs) with the perception that patients with SCD are addicted to pain medications; many HCPs have negative attitudes towards patients suffering with SCD (Freiermuth et al., 2014; Institute of Medicine, 2012). HCPs also delay pain medication administration and judge patients who are in need of understanding and receiving adequate pain medication administration (Institute of Medicine, 2012). The case manager can be a strong advocate for the patient and help educate the healthcare team to minimize negative attitudes and increase the likelihood of delivery of best practices (Case Management Society of America, 2016).

Thus, caring for persons with SCD is complex from both a medical and health services use perspective. There are many barriers to care. These barriers include: clinician and patient knowledge deficit, chronic pain and the perception of addiction, frequent visits, SCD stigma, and implicit bias. Identification and discussion of these barriers provide a framework for the case manager in a variety of healthcare settings that care for persons with SCD.

## BARRIERS

### Clinician and Patient Knowledge Deficit

Comprehensive educated and coordinated care for the complex and disabling effects of SCD is necessary for successful patient outcomes. Despite the integral role of healthcare providers and patient self-care, studies demonstrate that lack of provider knowledge and negative provider attitudes are significant barriers to care (C. Haywood, Jr. et al., 2009). Most primary care physicians have limited experience caring for patients with SCD, and few have more than five patients with SCD (Mainous et al., 2015). Individualized discharge plans with direct patient involvement can reduce barriers faced after discharge and reduce readmission rates (Brodsky et al., 2017; Cakir, Kaltsounis, K, Kopf, & Steiner, 2017).

Effective management and coordination of advanced therapies such as blood transfusion, hydroxyurea (HYDREA, DROXIA), and prophylactic antibiotics positively impact successful patient outcomes (Elmariah et al., 2014). However, providers are often uncomfortable managing patients with SCD who need blood transfusions, hydroxyurea and pain management (Mainous et al., 2015). Complex illnesses and complications of SCD impede primary HCPs' management of SCD as they often feel uneasy and incapable of knowing how to begin treatment.

Providers display increased difficulty managing SCD pain, and most do not agree that the self- report of patient's pain is the most reliable indicator (C. Haywood, Jr. et al., 2009). Nurses also report a lack of knowledge with narcotic analgesia use in pain crises and have limited experience with patients and SCD (C. Haywood et al., 2015). Recent studies have demonstrated improved provider attitudes toward patients with SCD after ongoing educational interventions (Freiermuth, Silva, Cline, & Tanabe, 2016; Hanik, Sackett, & Hartman, 2014). Case managers can provide real-time education to nurses and other healthcare providers that can lead to improved attitude and better pain management for patients with SCD. In 2014, the National Heart, Lung and Blood Institute (NHLBI) published evidence-based guidelines and recommendations for the treatment of VOE (National Heart, 2014). See Box 1. The case manager can alert staff to the guidelines that provide evidence on how to manage pain and other medical complications.

Self-care is an essential part of managing SCD. Patients must maintain medical appointments, keep up to date on immunizations throughout the lifespan, ensure adequate hydration, and pay special attention to changes in environmental factors. Identification of factors that place patients at risk for illness leads to greater success in SCD outcomes (Tanabe et al., 2010). Patients who lack the ability to maneuver through the healthcare system or have limited knowledge of their disease may not receive adequate pain control and suffer poor healthcare experiences and outcomes. The case manager can play an important role in assessing the patient's knowledge of the disease and self-management strategies. This knowledge may be beneficial for the patient's interactions with staff and for overall patient medical management. Advocacy is essential to the case manager's role, which includes putting the patient first, and knowing the patient's usual state of health and pain medication schedule (Tahan, 2016). The case manager can help the patient with SCD to navigate

transitions by promoting self-care, encouraging clear communication and educating the team about the patient's history.

## Chronic Pain and the Perception of Addiction

Many patients with SCD suffer from chronic pain and require long term opioids (W. R. Smith et al., 2015). Patients who suffer from chronic sickle cell pain present atypically and may not appear distressed when performing activities of daily living (Brown, Weisberg, Balf-Soran, & Sledge, 2015). Unfortunately, this contributes to a distrust of the patient's self-report of pain when health care professionals are confronted with a sickle cell patient with VOE (Brown et al., 2015). Distrust of patients' reports about their painful VOEs can lead to inadequate or poorly controlled pain. Health care professionals may doubt reports of pain by patients in crisis and take this condition less seriously due to the attitudes and beliefs about addiction and concerns regarding drug-seeking behavior (Shavers, Bakos, & Sheppard, 2010). The perception that patients with SCD have higher rates of opioid addiction is not uncommon among health care providers (Freiermuth et al., 2014). There is no evidence to support the belief that patients with SCD are addicted to opiates, yet health care providers continually label SCD patients as addicted. These negative attitudes demonstrated by health care professionals interfere with proper pain management in SCD (C. Haywood, Jr. et al., 2009).

Acute pain, VOE is the hallmark symptom of SCD and also the most common reason for hospital admission (National Heart, 2014). Most patients try to treat the pain at home, but when the pain exceeds at home management, patients seek further medical care, but generally only after exhausting all pharmacological options (Adegbola et al., 2012; Aisiku et al., 2009; Booker, Blethyn, Wright, & Greenfield, 2006). VOE requires high doses of opioids, which often lead to the perception of addiction by healthcare providers. Pain is a subjective finding, making it difficult to objectively measure, thus, the patient's self report of pain may not be considered. Research demonstrates that SCD patients in severe pain wait significantly longer to be evaluated by a HCP and receive delays in much needed pain medications (C. Haywood, Jr. et al., 2014; Tanabe et al., 2007). Case managers can advocate for the patient and educate the healthcare team on misperceptions of addiction, the patient's usual levels of pain and the complexity of caring for patients with SCD may lead to improved patient care. Case managers have an important opportunity to take the lead on identifying the factors that impede quality and safe care.

## Frequent Visits

Another important barrier to care for persons with SCD is being a frequent healthcare visitor. Patients with SCD have more frequent hospitalizations than other patients with chronic medical conditions (Jiang et al., 2014). Patients with asthma, pneumonia, heart failure and diabetes mellitus have lower 30 day rehospitalization rates than patients with SCD, and one in three SCD patients are readmitted within 30 days (Brousseau, Owens, Masso, Panepinto, & Steiner, 2010). There are a small number of SCD patients who have a significantly high hospitalization rate, 4 or more hospitalizations per year, and also have longer length of stays with higher readmission rates than other Medicaid users (Jiang et al., 2014). Although this number is small, because they have a large number of visits, there is a

generalization that all patients with SCD are frequent patients and addicted to opioids (Freiermuth et al., 2014).

Poor provider attitudes are associated with SCD patients who have frequent hospital visits (Ratanawongsa et al., 2009). Those same poor provider attitudes can lead to patients that delay seeking treatment which may negatively impact their health care experience and outcomes (Jenerette, Pierre-Louis, Matthie, & Girardeau, 2015). Hospital admission and readmission rates are more complex for the individual with SCD because they require care coordination, complex medication regimens, transportation to and from day treatment centers, extensive social support and judicious follow up care (Cakir et al., 2017). The case manager can identify individual socioeconomic risk factors to hospitalization and rehospitalization, ensure access to adequate pain medications and schedule follow up provider appointments which can reduce overall costs and prevent future hospitalizations and readmissions for patients with SCD (Cakir et al., 2017).

The case manager can take an active role in team rounds by advocating for patients with SCD by reminding staff that patient vulnerability is increased with frequent admissions and call attention to negative attitudes and delayed pain medications. Case managers can also collaborate with all team members to coordinate effective discharge plans that include individualized pain management plans of care, and access to outpatient services to prevent recurring hospitalizations for SCD complications.

## SCD Stigma

Persons with sickle cell disease are subjected to stigma associated with their disease and are perceived as drug seeking, inevitably influencing their ability to receive quality care. The need for high doses of opioids and patient requests for specific pain medication dosages support the false beliefs that patients are drug seeking or addicted to pain medications. Nurses report increased levels of opioid addiction and greater frustration caring for patients with SCD compared with physicians (Freiermuth et al., 2014). Receiving pain medication within 60 minutes of arrival to the emergency department and use of individualized dosing protocols is recommended by the NHLBI guidelines (National Heart, 2014). Lack of adherence to these guidelines is closely associated with negative provider attitudes (Glassberg et al., 2013). Health care providers who are skeptical that patients are actually in acute pain or believe that they are addicted to pain meds leads to a decreased administration or hesitance to administer opioids (Freiermuth et al., 2014; C. Haywood, Jr. et al., 2009). This stigmatization causes SCD patients to experience longer wait times to see a physician upon arrival to an ED than do other patients with chronic conditions regardless of having presented with higher degrees of pain and a higher priority assignment in triage (C. Haywood, Jr., Tanabe, Naik, Beach, & Lanzkron, 2013).

SCD patients are well aware of this misperception and often feel mistrusted and stigmatized as a result (Booker et al., 2006). This further exacerbates the stigma and increases the barrier to receiving evidenced based care. Decreasing the stigma associated with sickle cell disease and advocating for patients is integral to the case managers' role. Case managers can work with healthcare providers by providing education about the disease and associated complex medical and social behavioral health needs, as well as the high risk of death.

### Implicit Bias

Both discrimination and unconscious bias persists in our health care system (FitzGerald & Hurst, 2017; James, 2017). Healthcare providers have implicit and explicit racial bias demonstrating positive attitudes towards white persons and negative attitudes towards persons of color (Hall et al., 2015). These biases may impact the daily management and lifespan of patients with SCD. The need for opioids as a primary treatment for SCD makes the SCD patient uniquely subject to racial bias as a patient population. Race negatively affects the delivery of equitable evidenced based care for patients with SCD with negative provider attitudes contributing to the disparity (Nelson & Hackman, 2013). Case managers can advocate for racial equity, educate peers and discuss the role of implicit racial unconscious bias in healthcare. The case manager can address disparities that help alleviate implicit bias as a barrier to satisfactory care for patients with SCD.

### Conclusion

Case managers are in the unique position to minimize barriers to care and foster improved health care outcomes for patients with SCD. Assessment and recognition of the barriers to care, collaboration and care coordination, evaluation and advocacy for comprehensive health care equity for patients with SCD is essential to the role of a case manager. Ensuring evidenced based care and facilitating delivery of equitable care may reduce hospital readmissions, and decrease health care utilization, thus improving health care outcomes for patients with SCD.

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**Box 1**

## Summary of Vaso-Occlusive Episode (VOE) Recommendations

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| Consider causes of pain other than VOE. |
| Conduct a thorough assessment of pain, including patient's recent analgesic use (opioid and non-opiod) as well as patient's knowledge of what agents have been successful in the past. |
| Initiate analgesic therapy within 30 minutes of triage or 60 minutes of registration (emergency department management). |
| Use an individualized prescribing and monitoring protocol, or SCD specific protocol to promote rapid, effective and safe analgesic management and resolution of VOE. |
| Treat with non-steroidal anti-inflammatory agents (NSAIDS) for persons with mild to moderate pain who report relief with NSAIDS, in the absence of contraindications. |
| For severe pain, rapidly initiate parenteral opioids. Calculate the parenteral opioid dose based on daily short acting opioids. |
| Administer opioids using the subcutaneous route when intravenous access is difficult. |
| Re-assess and re-administer opioids for continued severe pain every 15-30 minutes until pain is under control per patient report. |
| Initiate around the clock opioids by patient controlled analgesia (PCA) or frequently scheduled doses vs. "prn". |
| Do not use meperidine unless it is the only effective opioid for an individual patient. |
| For patients that require antihistamine for itching caused by opioid administration, prescribe orally. Re-administer ever 4-6 hours if needed. |
| Encourage the use of the incentive spirometer to prevent acute chest syndrome in hospitalized adults and children. |
| In euvolemic patients who are unable to drink fluids, administer intravenous hydration at no more than maintenance rate to avoid over-hydration. |
| Use an objective sedation scale to monitor for excessive sedation. |
| Gradually titrate down parenteral opioids as VOE resolves. |
| Do not administer blood for the treatment of VOE. |
| Administer oxygen for patients with an oxygen saturation <95% on room air. |
| Encourage use of non-pharmacologic approaches to manage pain including heat and distraction. |

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