Sickle Cell Disease Related Pain: Crisis and Conflict

Knox H. Todd, Carmen Green, Vence L. Bonham, Jr., Carlton Haywood, Jr., and Evera Ivy

Editor’s Note: This article is the second in a series of “Case Studies in Pain” features, designed to share scientific and clinical knowledge in a case study format. This report presents the case of a 17-year-old male treated for severe pain related to sickle cell crisis.

Case Study

A 17-year-old African American male was admitted to the Emergency Department (ED) for complaints of severe pain related to sickle cell crisis. He reported to the ED physician that he usually required hydromorphone (Dilaudid) 5 mg intravenously when the pain was this severe. He also requested diphenhydramine 50 mg as the opioid usually caused pruritus. An IV was started to provide hydration.

As the nurse began to administer the medications, the patient requested they be given in the port closest to the insertion site and that they be pushed rapidly. In 1 h, the patient requested another dose of both drugs, despite appearing to be quite sedated. When the nurse expressed reluctance to give the dose in the manner he requested, the patient became upset.

Case provided by Judith A. Paice, PhD, RN
Director, Cancer Pain Program
Division of Hematology-Oncology
Northwestern University
Feinberg School of Medicine
Chicago, IL

Best Practices in the Emergency Department

Almost all of the 75,000 annual hospitalizations for vaso-occlusive pain crises related to sickle cell disease involve treatment in the ED.17 ED care for this population of patients is challenging as the emergency physician often has little knowledge of the patient’s prior medical history, particularly when the patient has no identifiable primary care provider or a history of unsatisfactory relationships with multiple providers. Patients with sickle cell disease may have experienced a lifetime of recurrent pain crises with inconsistent and often inadequate treatment, related to a host of medical, economic, and social factors.

Conventional therapies for acute vaso-occlusive pain crises involve oxygen, fluid replacement, and analgesic administration as well as specific interventions targeted to potential precipitating factors, such as infection. The evidence to support oxygen and fluid replacement interventions is limited. Prospective randomized, controlled clinical trials have demonstrated that 50% oxygen administration is associated with significant reductions in the number of reversibly sickled red blood cells, compared to subjects treated with room air. Unfortunately, this reduction is not associated with the duration of pain crises, hospital admission rates, or analgesic needs.26,35 Thus, there is no evidence that supplemental oxygen therapy alters outcomes in patients with normal arterial oxygen saturations. On the other hand, oxygen is readily available and inexpensive, thus, is commonly used in this setting. Intravenous fluid therapy is another commonly employed intervention that is supported by very little, if any, scientific evidence. Theoretically, increases in intravascular volume should limit vascular sludging in capillary beds, thus reducing pain. For the euvolemic or mildly hypovolemic patient, oral hydration is an appropriate choice. Whether intravenous volume administration was needed in this case is unclear.

Although acetaminophen and nonsteroidal anti-inflammatory agents are indicated for almost all patients with pain, opioids also will be required for virtually all patients presenting to the ED. Opioid therapy should be aggressive and many patients with pain crises will require greater doses than the general population due to high pain intensity, accelerated renal clearance and hepatic metabolism in some patients, and possible pharmacologic tolerance.4 In EDs providing high-quality pain
Sickle Cell Disease Related Pain

The clinician’s responsibility in this case is to treat the patient’s pain with compassion and avoid causing harm. In this case, the patient requests a relatively large dose of a specific opioid analgesic, hydromorphone. He requests that it be delivered in a fashion that will result in a rapid rise in serum levels with an additional request for co-administered diphenhydramine. Additional requests for hydromorphone are met with reluctance and the patient becomes angry. We are not told, but it is likely, that the ED staff has no contact with the patient’s continuity physician, if one indeed exists. Presentations similar to this are a common occurrence in the ED.

The clinician’s concern in this case is that the patient is seeking analgesics for reasons other than those strictly related to the relief of pain resulting from vaso-occlusive crisis. Generally, there is no method to assess underlying vaso-occlusive pathophysiologic alterations directly, and the patient must be assumed to have pain of physiologic origin.

The patient’s actions are perhaps best interpreted as “aberrant drug-related behaviors,” and there is an extended differential diagnosis for such behaviors the clinician should consider. Such behaviors may be manifestations of addiction, involving out-of-control behavior, or compulsive, harmful drug use; however, it is unlikely that sufficient information will be available during a single ED encounter to make this diagnosis. It is possible that these requests represent a form of pseudoaddiction—perhaps the patient has previously received large doses of hydromorphone in combination with diphenhydramine, delivered in a rapid intravenous bolus with resultant rapid relief of pain and simultaneous euphoric sensations. This experience may have contrasted favorably to past approaches resulting in delayed and insufficient pain relief. To the extent that this treatment regimen was successful in relieving pain, the patient would understandably request similar treatment.

It is possible that the patient is seeking relief from multiple symptoms in addition to pain, such as anxiety or depression related to an underlying mood disorder. Chronic pain is often accompanied by mood disorders and psychiatric comorbidities that complicate the management of these challenging patients. The presence of aberrant drug-related behaviors in patients with borderline personality disorders may represent an expression of fear and anger or an attempt to cope with chronic boredom. Such patients may use opioids and alcohol in attempts to lessen symptoms of anxiety, panic disorder, depression, or insomnia. In obtaining a thorough history one might elicit such clues; however, this is unlikely to be achieved in the midst of a pain crisis and is best deferred to a later time. Emergency physicians often receive limited training in dealing with psychosocial disorders and our specialty’s deficiencies in dealing with such problems have been documented. After the immediate crisis passes, psychiatric consultation, if available, may be useful in both suggesting alternative causes for aberrant behaviors and tailoring a therapeutic approach to deal with these complicating factors. With an open, honest discussion of the clinical issues described above, it may be possible to achieve positive results. Without such a discussion, hostility and mutual medical mistrust is the predictable and all too common result.

Knox H. Todd, MD, MPH
Director, Pain and Emergency Medicine Institute
Department of Emergency Medicine
Beth Israel Medical Center
Albert Einstein College of Medicine
New York, NY

Racial and Ethnic Minorities and Disparities in Care

Sickle cell disease (an inherited disorder of the red blood cells) affects 1 in 600 African Americans. Although sickle cell disease is found in other ethnic groups (eg, Hispanics) where malaria is endemic, it is also the most common genetic disorder among people of African descent. Sickle cell disease is associated with a myriad of symptoms related to pain (eg, priapism, splenic infarction, avascular necrosis, dactylitis, acute chest syndrome, osteomyelitis, acute bone pain, stroke, susceptibility to infections) and is frequently characterized by acute painful crises due to vaso-occlusion. There is great variability in the frequency of painful crises. Although some people with sickle cell disease have mild painful episodes due to vaso-occlusion that readily respond to conservative treatment, others have severe painful episodes that require hospitalization and treatment in the ED, and still others have chronic persistent pain with acute exacerbations.

Painful episodes often occur without a precipitating event and may be short lived (ie, a few hours) but most painful episodes last almost a week. For those experiencing acutely painful crises, 60% have at least 1 severe episode a year with 20% experiencing at least 1 severe episode each month. The result is decreased work productivity, missed work, and school absences that significantly impair a patient’s social support network and quality of life. The relapsing and recurrent nature of sickle cell disease leads to all of the long-term sequelae associated with any chronic pain problem (eg, increased disability, depression, etc). Increased pain intensity and frequency of painful episodes results in increased morbidity, although diminishing quality of life and life expectancy.

Under treatment of pain is often related to issues of disparity. The pain complaints of racial and ethnic minorities are less likely to receive the attention that they deserve (particularly in the acute care setting). The gold standard for pain assessment remains patient report. The patient’s behavior, how patients talk about their pain, physician-patient communication, and stereotyping can complicate pain assessment. It follows that race and ethnicity are important in assessing pain. In...
addition, differences in the way that racial and ethnic minorities communicate their pain experiences (especially if there is a language barrier) as well as how physicians hear their concerns may lead to their pain complaints being discounted.29 Problematic communication is particularly likely if the patient’s gender, race, or ethnicity is not congruous with the physician’s.25,32,33

Pain education within the medical, nursing, dental, and pharmacy curricula is a neglected topic that contributes to poor pain assessment for all patients (especially racial and ethnic minority persons). Curricula directed at achieving cultural competence and classes that facilitate active listening among health-care professionals are also lacking. Physicians often have lesser goals for treating acute and chronic pain than for treating cancer pain. Nonetheless, health care professionals often report confidence in their pain management knowledge as well as in their ability to assess and treat pain. However, the literature supports that their confidence is often misplaced.

An important element when considering disparities in health is the diversity of the health care workforce. In an increasingly diversifying U.S. population, the majority (nearly 90%) of health-care professionals (ie, dentists, nurses, pharmacists, and physicians) are non-Hispanic white Americans, whereas nearly 30% of the U.S. population consists of African Americans, Native Americans, or Hispanic Americans. More specifically, only 8% of currently practicing health-care professionals are African American, Hispanic Americans, or Native Americans. These differences in the ethnic background of patients and health-care providers support the need to work at understanding people who are different from ourselves to provide care in a culturally sensitive manner, thereby improving the nation’s colloquial health.5

Beyond differences in race, health care providers and patients with sickle cell disease often differ based upon socioeconomic stratification. Although health insurance coverage provides access to medical care and improves overall health, even when sickle cell disease patients have a primary care physician and health insurance they often experience difficulty in securing referrals to pain medicine specialists. Individuals without health insurance or a primary care physician are more likely to use EDs for routine care. In addition, racial and ethnic minorities are less likely to have a regular primary care physician and therefore have less access to specialty care. The sickle cell patient presenting to the ED in an acute pain crisis is often where the health care provider-patient interaction for minorities and their health care professionals is fraught with the potential for racial stereotyping, mistrust, and problematic physician-patient communication.

In the middle of the Decade for Pain Research and Control, it is important to realize that sickle cell disease is a cause of undue suffering as well as acute and chronic pain for a large population in the U.S. (and across the world). Providing compassionate and appropriate analgesic care for sickle cell patients is consistent with pain relief being a human rights issue and the undertreatment of pain as fundamentally a medical error.

Carmen Green, MD
Associate Professor
University of Michigan Medical Center
Ann Arbor, MI

Legal, Ethical, and Social Considerations

The case identifies the patient as a 17-year-old African American male. What is the role of race, gender, and age in the context of assessment and treatment of this patient’s pain? A full appreciation of the legal and ethical tensions present in this case requires an understanding of the larger social context of sickle cell disease (SCD) in the U.S. Sickle cell disease has erroneously been characterized clinically and socially as a “black disease.”27 The connection of the disease with race has resulted in ethnic and social ramifications that are tied to race and racism in our society. Empirical evidence supports disparities in pain treatment based upon race and ethnicity.13 There is also evidence that the social experiences of black men adversely affect their experience with the United States health-care system.34 The issues of race and gender in pain treatment may influence the physician-patient communication and decision-making process.

The case does not state if the 17-year-old patient was unaccompanied by a legal guardian to the ED. As a minor under U.S. common law, parental consent generally is required for medical treatment of minor children. However, the legal system generally recognizes that most adolescents have the cognitive capacities to make appropriate health-care decisions, and the law has generally upheld physician’s assessments of these minors as “mature”.9 Common and statutory law generally supports health-care professionals in providing emergency care for children and adolescents in the ED without the consent of a parent or guardian.8 The American Academy of Pediatrics Policy Statement recommends that appropriate medical care for the pediatric patient with an urgent or emergent condition should never be withheld or delayed because of problems with obtaining consent.8 A sickle cell disease pain episode is an emergent condition.

With this background as context, what is the clinician’s responsibility when faced with this type of scenario? This case presents a tension between a clinician’s moral and legal obligation to relieve the pain of a suffering patient and moral and legal obligations to avoid the administration of unnecessary medications. Each of these obligations is supported by multiple ethical and legal sources. Ethically, these obligations are supported by health-care professional codes of ethics,1,2 the moral goals internal to medical practice6,24; and the ethical principles of beneficence (promoting the wellbeing of the patient), nonmaleficence (not inflicting harm upon a patient), and respect for autonomy, which among other things requires an acknowledgment of a person’s decision-making rights, require that clinicians assess the capacity of
the patient to participate in medical decision-making, and require that clinicians enable patients to participate in decisions concerning their care. The basis for these obligations are found in the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) guidelines and state medical board guidelines. JCAHO has established that all patients receiving care at a JCAHO-accredited institution have the right to the appropriate assessment and management of their pain. The Federation of State Medical Boards has developed model guidelines that recognize that opioids may be essential to the relief of acute or chronic pain, and that physicians ought not fear disciplinary action for the use of opioids for legitimate medical purposes. However, not all state medical boards have adapted their policies after these model guidelines, and some state medical board policies have in place regulations that can hinder a physician’s ability to use controlled substances for patient pain management.

The American Pain Society (APS) has published a “Guideline for the Management of Acute and Chronic Pain in Sickle Cell Disease,” which was the nation’s first evidence-based pain management guideline tailored specifically for SCD and is supported by an advisory group of the National Heart, Lung, and Blood Institute of the National Institutes of Health. The APS guidelines specifically speak to the issue of a sickle cell patient’s request for particular medications and dosages as illustrated in the case study. The APS guidelines recognize the fact that sickle cell patients typically are very knowledgeable concerning the medications and dosages that have worked for them in the past, so requests for specific medications at specific dosages alone should not be taken as an indicator of drug-seeking behavior. The short interval at which the patient in this case requests additional medications may be justified by considering that the patient’s prior experiences of pain and its treatment may have caused him to develop a tolerance to the medications, which is defined as “a state of adaptation in which exposure to a drug induces changes that result in a diminution of one or more of the drug’s effects over time.” The APS guidelines state that increasing dosages and shortening intervals between doses are both appropriate adjustments that may be made to overcome tolerance. The APS guidelines are not law but provide clinicians with guidelines based upon expert development. These guidelines would be introduced as evidence in support of a violation of the standard of care in any legal claim of malpractice.

Patients with SCD have been subject to a “skepticism” on the part of clinicians concerning the authenticity of the SCD patient’s report of pain. Studies of clinician attitudes have found that physicians and nurses both tend to greatly overestimate the prevalence of addiction in the SCD population, which in the literature is reported to range anywhere from 0 to 11%. These attitudes contribute to SCD patient reports that emergency room visits for their pain are often a dehumanizing experience.

The patient’s request in this case for additional medications at a short interval is not inconsistent with the standard of care for SCD and should not be taken without evidence as an indicator of a substance abuse problem. The clinician in this case should adhere to the patient’s request for medications in the manner requested. After administration of this dose, the clinician must continue to monitor the patient and assess his level of pain relief.

The reluctance on the part of the clinician in this case to provide the additional dose of medications in the manner requested by the patient expresses doubt concerning the authenticity of the patient’s pain. All patients have the legal and moral right to effective pain management. Should the clinicians in their continued assessment of the patient discover evidence that the patient is a substance abuser, a pain management/substance abuse specialist from the institution should be consulted. This specialist could help in the assessment of the patient’s substance abuse problem, as well as help in the development of a plan of pain management that could account for the patient’s substance abuse problem.

Vence L. Bonham, Jr., JD
National Human Genome Research Institute
National Institutes of Health
Bethesda, MD

Carlton Haywood, Jr., MA
The Phoebe R. Berman Bioethics Institute
Johns Hopkins University
Baltimore, MD

Personal Reflections

I was diagnosed with SCD as a child. I have been very fortunate in that I am generally hospitalized only once a year, and in fact, have had some years with no hospital admissions. I have about one to three pain episodes per month, but am usually able to manage these at home. If I notice that the pain is beginning, I take some acetaminophen, alone or with codeine, and get a good night’s rest. Usually I’m okay when I wake in the morning, unless there are other problems.

Like the patient in this case, when I have had to go to the ED, my experiences were generally not positive. Most of the time doctors and nurses didn’t trust that I was as sick as I said I was. They would not give me adequate doses of pain medications. My most recent hospitalization was 10 months ago. After several hours of trying to see if I could take care of my illness at home, I discovered it wasn’t possible because my crisis was getting worse. After getting to the emergency room, I was given 5 mg of morphine intravenously, which is a really low dosage. I had asked for Demerol, which seems to work better for me, but was told it was not available. Unfortunately, my primary care physician was out of town. It wasn’t until many hours later, after receiving the same ineffective dose, that they finally changed the medication.

When the doctors and nurses don’t trust me, and won’t give me adequate pain medications, the pain crisis becomes more severe. The longer and more severe the pain crisis, the longer recovery takes. So if I can go into the
hospital, receive good pain control, I would need treatment for 1 day. Instead, when pain control is inadequate, my crisis worsens, and my hospital stay is increased. This means I miss more school or work.

For example, because of ongoing pain and severe fatigue, I often had to miss a week or 2 of school, even when I was in the hospital only a day. It was very difficult to get back to school and to try to catch up on homework. It took me 6 years to get my bachelor’s degree. I was hospitalized my very first semester of college. I got sick after the date in which I could drop a class, which meant I received failing grades because of the work missed. I learned very early that, whereas some instructors were considerate and compassionate, not all were. I really questioned whether college was worth it and was ready to give up several times. It was during my final 2 years of college that I realized that the disease made me stronger and made me a better person.

I definitely feel that race (ethnicity) does play some part in the way we are treated. Although I do believe that there are some people who have the disease who have become addicted to drugs, it is not true for the vast majority of us. I have a college degree. I have been employed full-time since I graduated from college. I am a homeowner. I am working just as hard as everyone else. So to come into a hospital once every other year and to be stereotyped as someone wanting drugs is hurtful. I want nothing more than to be away from the hospital. I want to be at work. I want to be at home with my family. I want to be on vacation lying on a warm beach. I want to just enjoy my life.

Evera Ivy
Volunteer
Sickle Cell Disease Association of Illinois (SCDAI)
Chicago, IL

References

16. JCAHO: Joint Commission Focuses on Pain Management: JCAHO, 2005, 7-25-2005

27. Rotimi CN: Are medical and nonmedical uses of large-scale genomic markers conflating genetics and “race”? Nat Genet 36:S43-S47, 2004


