

Resolving Conflicts:

Misconceptions and Myths in the Care of the Patient with Sickle Cell Disease

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Abstract

Sickle cell disease is an autosomal recessive disease that primarily affects persons of African ancestry. The hallmark of the disease is hemolytic anemia and vaso-occlusive crisis. Patients often have recurrent and severely painful episodes that necessitate the use of opioids. The reluctance of some health care providers to prescribe narcotics has resulted in adversarial relationships with some patients. The socio-cultural disparity between patients and providers may play a role. However, the lack of knowledge and understanding of the underlying pathophysiology of the disease and pain are the key issues. Education, research and hands-on experience, resulting in changes in attitudes and behaviors, will ultimately lead to a more empathic approach to the sickle cell patient. **Key Words:** Sickle cell disease, opioids, vaso-occlusive crisis.

Background

SICKLE CELL DISEASE is an inherited disorder that is characterized by altered amino acid composition of the globin chain. While the presence of sickle cell disease most commonly affects persons of African descent, it can also occur in people from the Mediterranean region. The gene frequency in African Americans is about 8%, but may be as high as 50% in equatorial Africa. Approximately 1 in 500 African Americans is affected with a sickle hemoglobinopathy. Currently, 220 patients with sickle cell disease (150 children and 70 adults) receive their care at the Mount Sinai Medical Center. Of these, 80% are African and Caribbean American, and about 20% Latino.

Hemolytic anemia and vaso-occlusion are the hallmarks of this disorder. Vaso-occlusive events

may occur acutely or may be recurrent. These events can be quite frequent and severe, and when they occur in vital organs they can be life threatening. There can be considerable variability in the clinical course of different patients with sickle cell disease, and even—at different times—in the same patient.

A discussion of the mechanism and pathogenesis of vaso-occlusion is beyond the scope of this paper. However, there is increasing evidence that several factors, such as erythrocyte-endothelial interactions, vascular modulation, as well as hemostasis may play a role in the vaso-occlusive phenomenon (1). Genetic modifiers such as co-inheritance of thalassemia and fetal hemoglobin expression may also influence clinical expression.

Discussion

The acute, painful “crisis” accounts for most of the hospital visits of adult sickle cell patients. Because of the risk of overwhelming infections in the pediatric patients, episodes of fever account for a considerable number of pediatric visits. In most patients, there is no obvious precipitating factor for a painful crisis. In some patients, infec-

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tion, extremes in temperature, and physical or emotional stress may precipitate a crisis.

The pain typically involves the back, extremities, chest and abdomen but may occur anywhere. It is important to note that, in many cases, pain can and does occur without objective physical signs. The severity and frequency of pain can vary considerably among patients and sometimes in the same patient. Interestingly, the pain rate (i.e., the numbers of episodes per year) has been shown to be a good measure of clinical severity and a predictor of early death in patients over 20 years old (2). About a third of all patients with sickle cell disease have recurrent vaso-occlusive crisis, and only a very small percentage (5–10%) of patients experience more than 10–20 events per year (3). Although acute, recurrent, painful episodes are often the predominant feature of this disease, it should be underscored that sickle cell disease is not a one-dimensional disease. The frequent need to use narcotics to treat severe pain often leads to drug dependence in some patients which results in their stigmatization by some health care professionals. What role, if any, do these factors play in cultivating misconceptions, myths, misallocations and distrust?

The acute painful episode is frequently the principal complaint that leads the sickle cell patient to interface with the health care system. Perhaps the best approach to understanding the origins of these myths and misconceptions, stereotyping and stigmatization would be to focus on the issues of pain and pain management. It might be helpful to examine certain elements that may be a source of the friction with this particular patient group.

Both the patient and health care provider must understand the management of sickle cell pain. Negative attitudes about narcotics and fears of addiction may bias both providers and patients, which results in the failure to use opioids appropriately. In addition, patients may not have the skills needed to cope with a chronic, incurable disease. Feelings of helplessness, which often surface in early childhood or adolescent years, can lead to maladaptive behavior. This is frequently manifested during encounters with the health care system. Several fundamental questions should be addressed:

- What is the knowledge and understanding of sickle cell disease among health care providers?
- Do health care providers have a basic understanding of the pain of sickle cell disease?
- What is the patient's understanding of his/her disease?

- Since this is a disease that affects the entire family and support system of the patient, does the health care provider understand the psychosocial background of the patient?

In this discussion, we will attempt to address how and why these issues may impact on the provision of adequate and effective treatment for this group of patients, and propose strategies that may assist the medical community in developing a more candid relationship with the sickle cell patient.

Understanding the Pain of Sickle Cell Disease

Health care providers often fail to appreciate that there are different types of pain syndromes that occur in patients with sickle cell disease. Table 1 lists the different types of pain that a sickle cell patient might experience (4). It is important to recognize and appreciate these differences, because acute painful episodes are unpredictable in onset and severity.

Sickle Cell Pain

Many health care providers mistakenly believe that all pain is the same and, therefore, can be treated the same. But, pain research focused on the neurobiology of pain has established that the pathophysiology of pain in sickle cell disease is quite different from that of other pain syndromes. Ballas et al. have discussed this in detail (4). The standards and guidelines in the management of pain in patients with cancer and other pain syndromes are well established and have been widely published. These guidelines are helpful tools in the management and appropriate

TABLE 1

Types of Sickle Cell Pain

Acute	Painful crisis
	Acute chest syndrome
	Right upper quadrant pain
	Cholecystitis
	Hepatitis
	Priapism
	Dactylitis (Hand Foot Syndrome)
	Splenic sequestration
Chronic	Avascular necrosis (hips/shoulder)
	Vertebral collapse
	Leg ulcer
	Arthropathy
	Arthritis

use of opioids and other analgesics for chronic pain of a finite duration. They do not, however, address the issue of the recurrent or chronic pain that is unique to sickle cell disease. The frequent, recurring and often chronic pain experienced by sickle cell patients sets it apart from many other pain syndromes in that this disease is lifelong and for most patients incurable (5).

Believe the Patient

The report of pain by any patient must be considered accurate. If the patient believes that he or she is not being taken seriously, and the physician is suspicious of the patient's motives in seeking care, an adversarial relationship is quickly formed.

Although this principle of trust in one's patient is crucial, it does not exclude a need for caution. Because the pain that some patients experience is chronic, the misuse of opioids does become a potential risk. Tolerance and sometimes dependency can become real problems. One contributing factor may be the lack of understanding of pain management by both the provider and the patient. To circumvent these problems, the use of adjuvant non-opioid analgesics, pain diaries, patient care contracts and non-pharmacologic methods (relaxation and diversion methods) have proven helpful for some patients. Patients who exhibit problem behaviors and those who tend to overutilize emergency room services should be identified. It is essential that each patient be evaluated and assessed on an individual basis.

Failure to Assess Pain Accurately

Accurate pain assessment is critical for adequate treatment of acute and chronic pain. Improper assessment of pain and failure to monitor the response to therapy are major factors in the failure to relieve pain. Patients are very different from one another in their clinical presentations. A patient who has infrequent painful episodes and who is considered "opioid naive" may require much less analgesic to relieve pain than someone with frequent episodes, who might be "opioid tolerant." Pain scales that allow a thorough evaluation and assessment of pain should be used; they may help to alleviate conflicts over appropriate dosage.

"Good Sickler"/"Bad Sickler"

For many patients with sickle cell disease, pain is part of their daily lives. This can indeed

lead to misuse of the opioid therapy prescribed. Such misuse, in turn, can lead to tolerance, dependency and overutilization of hospital services by a very small subset of sickle cell patients. This group of patients generally accounts for many hospital emergency room visits. These patients, often labeled "drug seekers," can induce the staff to form false beliefs about all sickle cell patients. Such stereotyping interferes with the adequate and effective treatment of pain for other sickle cell patients. The failure by health care providers to distinguish between addiction, dependence and tolerance is a major component in the failure of effective management of the sickle cell patient with pain. It can lead to conflict and adversarial relationships.

Strategies to Improve the Provider-Patient Relationship

Most hospitals subscribe to and display a patient's bill of rights. Recognizing a patient's rights and clarifying patient responsibilities are paramount for improving the provider-patient relationship.

Patients have the right to:

- Considerate and respectful care.
- Confidentiality and privacy.
- Current information regarding their management.
- Participate in the planning of their care (unless it is judged unsafe for them to do so).
- Informed consent.
- Refuse treatment.
- Voice grievances about care or services.
- Continuity of care.

What role does the patient play in helping to dispel the myths and misconceptions about patients with sickle cell disease? A relationship that is grounded in mutual respect requires balanced input and therefore obligates the patient to uphold certain responsibilities.

Patients' responsibilities include:

- Providing, wherever possible, accurate and complete information about their symptoms and medications to the various health care workers involved in their care.
- Complying with instructions given related to care and medications provided.
- Asking questions and participating in the planning of their care.
- Understanding the management of pain and the careful use of opioids and analgesics.

- Understanding the importance of establishing a trusting relationship with their provider (4).

Changing Attitudes and Behaviors

As health care providers, our attitudes and beliefs influence the relationships that we form with our patients. These beliefs affect the care and management we provide. Attitudes and beliefs about pain and pain management, particularly the use of opioids, are typically negative. Denial of the importance or presence of pain and fears of addiction are the basis for this bias.

How do we change attitudes and behaviors? One obvious answer is education. Curricula for physicians, nurses and other health care professionals have been developed by the International Association for the Study of Pain, and teaching materials are available (6). Table 2 lists several factors that affect the use of analgesic medications and possible strategies to change behaviors.

The Difficult Patient

As previously mentioned, a small subset of patients is difficult to manage and triggers negative reactions in some health care providers. Possible solutions for dealing with these patients include:

- Providing continuity of care by designating a single physician or health care team.
- Offering psychosocial support (i.e., family, other patients, support groups and psychiatric intervention).
- Designing patient care contracts that set limits.
- Designating a single pharmacy for prescriptions.
- Being consistent with the plan of care.

Conclusion

Sickle cell disease is an inherited chronic illness that carries the risk of significant morbidity and mortality. Lack of knowledge about sickle cell disease and the difficult issues of pain management lead to negative attitudes among providers toward patients with the disease. Through research and education, changing attitudes and behaviors will lead to establishing a

TABLE 2

Factors Affecting the Prescription of Analgesic Medications

Factors	Corrective Strategy
Denial of the existence of pain	Educate; change policies to require measurement of quality assurance of pain; file complaints.
Denial of the importance of pain	Monitor and record pain; educate about long-term effects of pain.
Unfamiliarity with effective use of analgesics	Pain management education.
Fear of adverse effects (e.g., respiratory depression and addiction)	Educate about extent and management of side effects.
Fear of scrutiny by regulatory agencies	Ensure that regulatory agencies are sensitive to management.
Family views	Examine underlying anxieties; give information.
Patient/family fear of tolerance, addiction and drug abuse	Patient/family education directed at false beliefs.

more reciprocal relationship, and thus more effective management of the patient with sickle cell disease.

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