NEW! **CASE STUDIES IN** PAIN MANAGEMENT

Episodic Pain in Sickle Cell Disease

In this first article of a new series, the authors describe a patient with acute lower extremity pain due to sickle cell disease and make their case for a more aggressive therapeutic approach to this type of pain.

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Sickle cell disease (SCD) is a set of genetic abnormalities primarily affecting patients of African and Mediterranean descent. In the United States, approximately 80,000 people currently have SCD and about 1000 babies are born with the disease each year. Because the disease is relatively uncommon, emergency department management is highly variable from institution to institution and represents a significant challenge for many clinicians.

Pain is the primary symptom of SCD, and severe, paroxysmal, painful vaso-occlusive episodes, often inaccurately called "crises" (see box on page 9) are the most common reason for presentation to the emergency department. Acute sickle cell pain has been described as worse than postoperative pain and as intense as cancer pain. The pain most frequently involves the abdomen, chest, back, and extremities. Episodes can be precipitated by infection, dehydration, extremes of temperature, changes in altitude, and stress, or they may have no clear trigger. Most patients have their first painful episode by age two. Some patients visit the emergency department two or three times a year for pain, but they are in the minority.

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The pathophysiology of pain is complex and even more so in chronic pain disorders.

The following case study describes a patient with SCD who is well known to the health care system and has multiple visits to the emergency department each year.

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PATIENT PRESENTATION

Mrs. R, a 36-year-old African-American with a history significant for SCD (S beta-thalassemia) presents to the emergency department with acute bilateral lower extremity pain that woke her up during the night. The pain was unrelieved with her usual home dose of 15 mg of morphine. She says she was discharged from the hospital seven days ago for sickle cell pain and that this current episode is similar in intensity.

A review of symptoms is positive for chest pain, dyspnea, and vomiting. Mrs. R's medical history includes acute chest syndrome, mitral valve prolapse, pelvic inflammatory disease, and intermittent blood transfusions. Her medications are 1 mg folic acid daily, 15 mg morphine sulfate every 12 hours, 25 mg promethazine as needed, 1500 mg hydroxyurea daily, two tablets acetaminophen/oxycodone 325/5 mg every 4 hours (as needed), and one injection of medroxyprogesterone acetate every three months.

The physical examination reveals bilateral lower extremity tenderness from hip to ankle, a systolic ejection murmur, and tachycardia. Chest films are negative. Laboratory values show a reticulocyte count of 3.6%, a blood glucose level of 297 mg/dl, a serum potassium level of 3.4 mmol/L, and a hemoglobin level of 6.8 gm/dl. (The patient's baseline hemoglobin level was 10 to 11, with her most recent level on discharge at 9.)

Mrs. R is managed with one 5-mg dose of IV morphine, followed by three 10-mg doses at varying intervals (from 30 to 90 minutes), for a total of 35 mg. Pain management is established by a preexisting acute management protocol individualized

to the patient. Mrs. R is admitted to the clinical decision unit for treatment of sickle cell "crisis" and anemia. There, she receives a unit of packed red blood cells and 10 mg of IV morphine. Before discharge, she is restarted on her usual oral pain regimen. She is discharged after 24 hours and advised to keep in close touch with her primary care physician and case worker.

Abandoning the Term "Crisis"

Pain is a complex combination of biological, psychological, and social stimuli or characteristics, which combine within episodes or vary between patients to produce different experiences and responses. Our data and that of other researchers show that ischemic pain from SCD and the underlying vaso-occlusive cascade associated with this pain are not sudden, present-or-absent phenomena. Clearly, pain in SCD is often chronic. The time has come for both the pain research and sickle cell research communities to collaborate more closely to reduce or eliminate SCD pain, the most morbid and common symptom of the disease. Accuracy in terminology when describing SCD is also critical. Abandoning the term "crisis"—an inaccurate, ill-defined research concept—is an important first step.

DISCUSSION

Mrs. R is typical of someone with SCD who is well known to the health care system. She makes multiple visits to the emergency department each year and is followed closely by the sickle cell outpatient team and a case worker.

The pathophysiology of pain is complex and even more so in chronic pain disorders. Sickle cell pain is chronic, and over time the disease has devastating systemic manifestations that result in a significantly shorter life expectancy. One-third of patients at our institution with SCD visit the emergency department three or more times a year, despite aggressive outpatient therapy and support. These patients have a higher clinical disease severity, more pain days, more home opioid use, and worse physical function compared to SCD patients with fewer emergency department visits. Recent data from our Pain in Sickle Cell Epidemiology Study suggest that hospital utilization counts (emergency department visits and hospital admissions) may underestimate pain by excluding episodes that are of short duration or are self-treated. Our data illustrate that pain may still be intense and debilitating on days when patients do not seek medical help.

Opioid analgesics are the mainstay of therapy for SCD pain. Opioids are most often administered on a short-term basis, usually parenterally, during acute care in emergency departments or hospitals. They may be continued orally for short periods, usually days to weeks, after a medical contact. However, opioids may also be used to manage pain recurrently or chronically for long periods by primary care physicians (PCPs) or SCD specialists. They may be prescribed in these ambulatory settings by physicians with varying degrees of opioid phobia and different levels of experience with SCD. In any case, by adulthood, most SCD patients have had more than 15 years of at least intermittent exposure to opioid analgesics.

Our position is that opioids are underprescribed for chronic pain. Those of us who adhere to this philosophy believe that:

- Physicians worry unnecessarily about legal retribution for opioid prescription.
- Physicians worry unnecessarily that patients will develop pathological opioid addictions with chronic exposure.
- It is unethical to undertreat patients with organic causes for their severe pain.
- Opioids do not cause tolerance if pain is present.
- Only opioids can successfully treat significant pain.
- Central sensitization explains pain in the absence of objective findings.
- Patients who complain of pain and want opioid therapy will benefit from receiving it.
- Some patients require extremely high doses to control pain.
- If the patient becomes physically dependent on opioids from chronic prescription but is not psychologically dependent, then this dependence is not a problem.

Emergency physicians need to recognize that pain is a significant component of the SCD process and should be treated as a chronic pain syndrome with

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frequent exacerbations. These exacerbations may be markers of disease severity and should be treated aggressively. We recommend using individualized care plans with involvement of the patient's outpatient physician, a clinical decision unit when available, and patient-controlled analgesia pumps when possible. We hope that significant research into SCD pain and acute and chronic pain management in general will lead to alternative pain therapies in the future.

Mrs. R was successfully managed utilizing a combination of an observation unit and a predefined pain management plan for her acute exacerbations that was developed with the assistance of her PCP. Her case is a good example of how a coordinated effort between the emergency department and the patient's PCP can help alleviate opioid-prescribing fears while managing SCD pain effectively. □

SUGGESTED READING

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