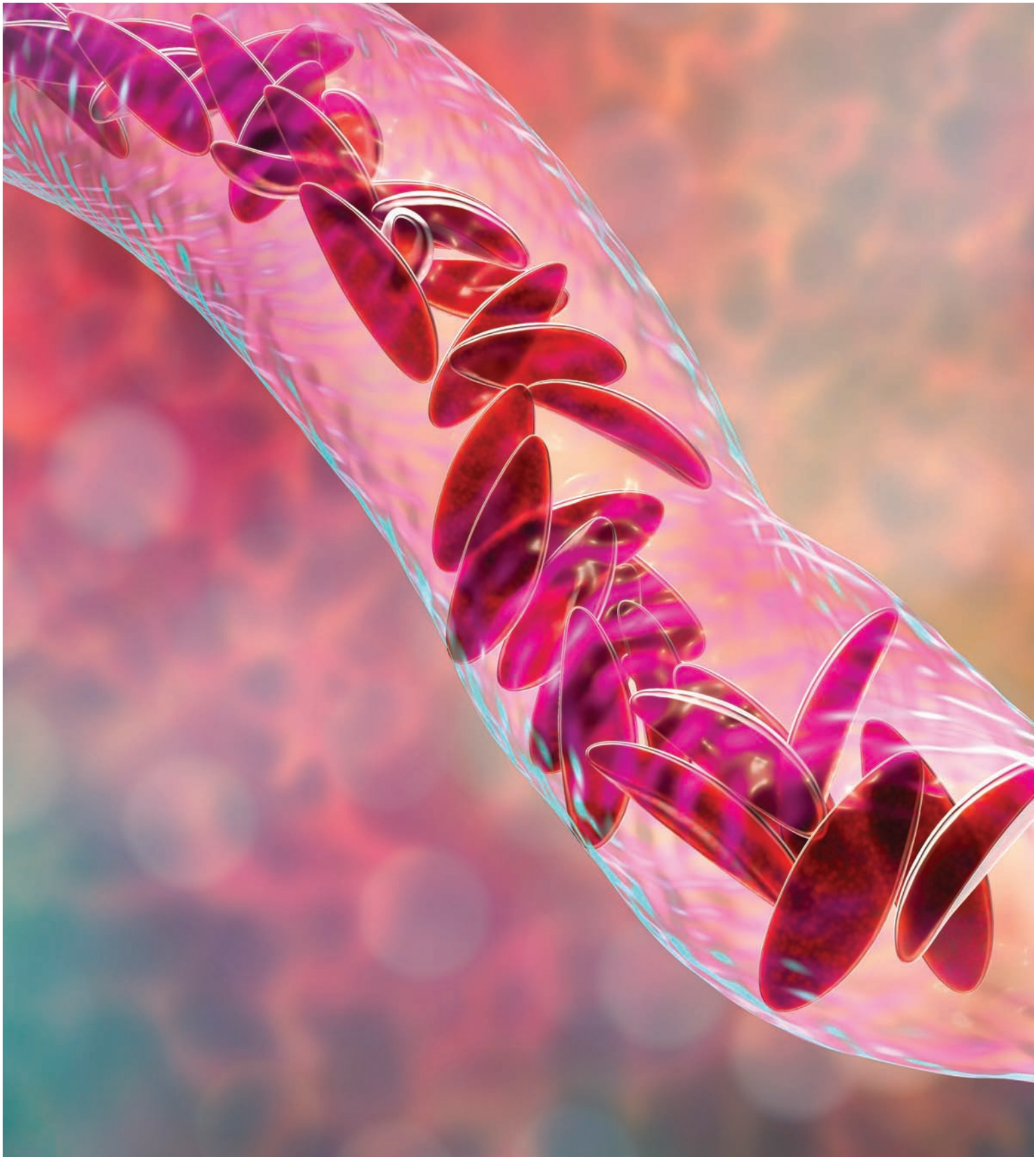

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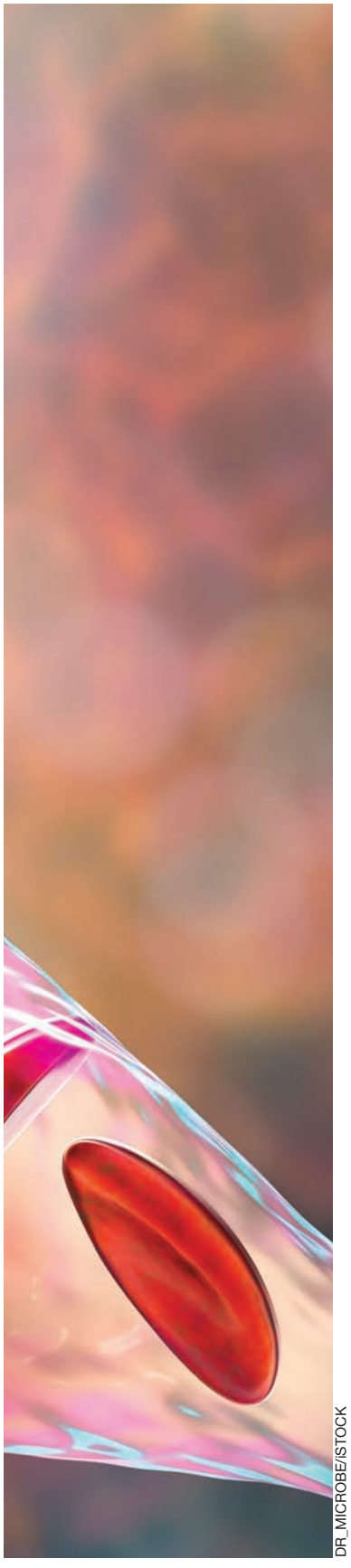

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Sickle cell anemia:

Best practices for patient-centered care

What are the treatment recommendations for this disorder that predominantly affects Black Americans? Follow along with a case study to learn how to minimize barriers and optimize outcomes.

By Kiernan Riley, BSN, RN; Michael M. Evans, PhD, MSEd, RN, ACNS, CMSRN, CNE; Kaléi Kowalchik, BSN, RN; Lucy Adams; and Megan Lucey

Sickle cell anemia (SCA) is an inherited blood disorder that causes the hemoglobin molecules in red blood cells (RBCs) to be defective. This causes the blood cells to have shorter lifespans and block blood vessels, resulting in anemia, fatigue, inadequate blood supply, and painful vaso-occlusive crises. Affecting approximately 100,000 individuals in the US, SCA has an estimated annual medical care cost of $1.1 billion. In the US, SCA predominately affects Black Americans, with an estimated 1 out of 365 births diagnosed with the disease, according to the CDC.

People of color often face implicit bias from healthcare providers and may be viewed with a more negative attitude than other patients. In addition, people with SCA may be viewed as "drug-seeking" because opioids are the treatment for the symptom of severe pain. The intensified stigma from the rising opioid crisis can result in pain not being managed properly and decreased quality of care for patients with SCA.

To address the complex issues surrounding treatment of SCA, a hypothetical case study is presented, which describes the events of a patient with SCA admitted to a medical-surgical unit following a vaso-occlusive crisis. A literature review was conducted using the scholarly databases CINAHL and PubMed to find the best practices for management of SCA in adults, including more education for healthcare professionals, utilization of alternative treatments, enhanced medical management, and better care coordination.

Case study

Mr. E, a 22-year-old Black male, presents to the ED with severe pain in his hands and abdomen. Mr. E has a history of anemia, gastroesophageal reflux disease, SCA, hypertension, and depression. He's currently a student at a local university where he studies psychology and plays soccer. His current daily medications include lisinopril, 20 mg; esomeprazole, 20 mg; and escitalopram, 20 mg.

Mr. E.'s vital signs are temperature, 100.4°F (38°C); pulse, 110 beats/minute; respirations, 18 breaths/minute; BP, 162/94 mm Hg; and oxygen saturation, 96% on room air. He complains of sharp, stabbing pain of 8/10 in his abdomen and hands and

has 1+ edema in his hands. Mr. E's skin is pale but warm to touch. He's awake, alert, and oriented x 4 and denies any other complaints.

After evaluating Mr. E, both the ED physician and nurse believe that he's experiencing a vaso-occlusive crisis as a result of SCA. They feel that this could be related to dehydration caused by recent warm temperatures and playing soccer. After receiving orders from the ED physician, labs are drawn for a complete blood cell (CBC) count and a complete metabolic panel. In addition, a 20-gauge I.V. is inserted in Mr. E's left antecubital fossa and normal saline solution is started at 125 mL/h. Morphine 2 mg I.V. is given for pain by the ED nurse and Mr. E is admitted to the medical-surgical floor for further treatment.

Upon arrival to the medical-surgical unit, the nurse helps Mr. E get comfortable in bed and provides him with a urinal because an accurate measurement of intake and output is needed. His vital signs remain unchanged, except for a slight temperature decrease to 100.2°F (37.8°C). Labs reveal a white blood cell count of 12.3 mcL, a hemoglobin level of 10.2 g/dL, a hematocrit level of 36%, and a blood urea nitrogen level of 28 mmol/L, indicating a possible infection, anemia, and dehydration.

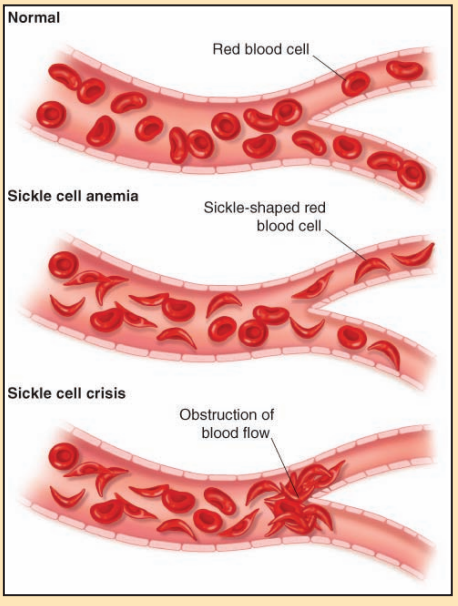
The hospitalist evaluates Mr. E and recommends that he continues his home medications and the I.V. normal saline solution at 125 mL/h. The hospitalist adds oral amoxicillin 500 mg every 8 hours due to Mr. E's fever, oral acetaminophen 650 mg every 6 hours as needed for a temperature greater than 100.4°F (38°C), oral folate 400 mcg daily, oral tramadol 50 mg twice a day, and a regular diet. The nurse provides Mr. E. with emotional support and he calls his parents to let them know what happened.

Pathophysiology

Predominately occurring in people of African, Arab, or Mediterranean heritage, SCA is a congenital hereditary disorder that affects the hemoglobin molecules in RBCs. In adults, the most common symptoms of SCA are vaso-occlusive crises, anemia, and hemolysis resulting in painful extremities. Other symptoms include fatigue, dactylitis (finger or toe inflammation), and jaundice. Patients with SCA have blood cells that contain an abnormal form of hemoglobin known as hemoglobin S in which both the normal beta-globin subunits of hemoglobin are replaced with hemoglobin S. This hemoglobin is defective, forming long rods that distort the RBC into a stiff sickle shape instead of its typical disc shape.

Due to their shape, sickled blood cells can't easily pass through blood vessels and get stuck, creating blockages (see *Picturing SCA*). Obstructed blood vessels lead to inadequate blood and oxygen supplies, which cause severe pain and injury to the affected areas, commonly referred to as a vaso-occlusive crisis. This crisis can last from several hours to days and may be caused by dehydration.

Picturing SCA



Source: Creason C. *Stedman's Medical Terminology: Steps to Success in Medical Language*. Philadelphia, PA: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2011.

Sickle-shaped blood cells are fragile and susceptible to rupturing, which releases hemoglobin into the bloodstream, causing hemolysis and hemolytic jaundice. These cells live for approximately 16 days, compared with healthy blood cells that live for an average of 120 days. A shortage of RBCs due to their sickle shape and short lifespan results in chronic anemia, which leads to fatigue.

Patients with SCA have an average hospital length of stay of 5.1 days, with an average cost per patient of over $7,500. In 2016, there were approximately 134,000 inpatient hospitalizations related to SCA. Approximately 90% of those hospitalizations were Black patients experiencing a pain crisis.

Barriers to care

Care of patients with SCA can be complicated, especially in the context of an acute vaso-occlusive crisis. Barriers to appropriate care exist, often stemming from social and system challenges. Some of the most commonly noted barriers include bias, stigma, healthcare provider availability and training, and inconvenient care.

As identified by Brennan-Cook and colleagues, being a person of color accessing the healthcare system can come with the impact of implicit bias of healthcare providers, which is generally viewed as a more negative attitude toward people of color than White patients. Compounding the possibility of implicit bias is the associated stigma of patients with SCA being "drug seekers." Darbari identified the intensification of this stigma following the rise of the opioid crisis. Because opioids are the standard treatment for SCA and one of the only interventions to reduce pain in this population, patients experiencing vaso-occlusive crises may have more difficulty obtaining opioids.

Beyond implicit bias and stigmatization, the healthcare system also creates barriers to appropriate care. Jacob and colleagues identified a lack of appropriately trained physicians to care for patients with SCA. Although many healthcare providers are aware of general protocols for SCA, research shows a current gap between implementation and utilization. Lack of clear universal guidelines and quick turnover rates create a barrier for seamless care of patients experiencing a vaso-occlusive crisis.

Patients may also experience extended wait times, which may cause a patient in crisis to use the ED for symptom control. However, the ED also presents distinct barriers, including overcrowding and undertraining of those providing care. Use of the ED also disrupts continuity of care and creates a disjointed trajectory of care for patients with SCA. It's within the nursing scope of practice to evaluate and advocate for our patients to overcome these barriers and enhance the use of best practices.

Best practices

According to the Expert Panel Report released by the National Heart, Lung, and Blood Institute, rapid analgesic administration of opioids, either I.V. or subcutaneous, is imperative and should begin within 30 minutes of the triage assessment in the ED to promote safe and effective relief of vaso-occulusive crisis. Doses should be increased by 25% until pain is controlled. Meperidine should be avoided in the treatment of vaso-occulusive pain. Nonpharmacologic treatments such as heat and distraction should also be implemented to help control pain.

did you know?

When a child inherits two sickle cell genes, one from each parent, he or she is born with SCA. If the child has one sickle cell gene, he or she will be a carrier of SCA but won't have the disease. Children with SCA will start to develop symptoms before their first year of life and are at risk for long-term complications, including anemia, vaso-occlusive crisis, splenic sequestration, stroke, infections, and priapism. Early diagnosis and proper maintenance of SCA in children are critical to prevent further complications and ensure that children have the greatest quality of life.

key points

Nursing considerations

- Spend time educating the patient and family about preventive and maintenance care to avoid an exacerbation of SCA, including seeing his or her healthcare provider at regular intervals and staying hydrated. Also, educate the patient about pain management and pain medications to ensure that he or she has the needed knowledge for proper self-care.
- Be mindful of the many psychological effects that SCA may have on the patient and coordinate his or her care accordingly. You may need to self-reflect about your attitudes toward pain management to ensure that you're creating a safe and open environment for the patient to receive care.
- Encourage supportive measures, such as heat, fluids, elevating affected joints, and opioids, to help manage and control the patient's symptoms.

Labs should be drawn stat and include a CBC, reticulocyte count, liver function tests, lactate dehydrogenase bilirubin, and electrolytes. In addition, patients with a pulse oximetry reading of less than 95% should be placed on supplemental oxygen and the use of incentive spirometry should be encouraged. Finally, I.V. fluids should be implemented at the maintenance rate based on the patient's weight to promote adequate hydration.

Due to the complex symptoms, stigmas, and financial burdens surrounding SCA, it can be difficult for patients to receive high-quality care throughout their disease process. Although there isn't a single best practice, the following practices can be implemented for patients living with SCA to better treat and manage their symptoms. With more education for healthcare providers on the pain and symptoms of SCA, exploration of both pharmacologic and nonpharmacologic interventions, and better screening and placement processes, treatment for patients living with SCA can become less fragmented, more person-centered, and deliver better outcomes.

Provision of nursing education on SCA

Regardless of the care setting, nurses are the main providers of direct care and play a critical role in pain management, patient education, and prevention of further symptoms or crises. Individuals with SCA have reported being stigmatized when seeking care for pain. These negative attitudes can be due to preconceived notions about pain and have the potential to act as a barrier between the nurse and patient. Pain assessment and treatment can be difficult because of the patient's subjective description of pain.

Studies show that negative attitudes regarding SCA pain extend past the ED and ICU and into medical-surgical nursing, so overall stigmatization needs to be minimized. Because nurses are the ones providing direct patient care, further education is crucial for nurses to better understand SCA pain so they can effectively educate patients. Research suggests that the potential for stigmatization because of inaccurate perceptions can be minimized by providing nurses with significant education about the pathophysiology and disease complications of SCA.

Use of complementary and alternative medicine therapies to treat SCA pain

Because of the current opioid epidemic, patients with SCA have reported decreased opioid dosing and increased stigmatization regarding opioid use. Although patients experiencing vaso-occlusive crises who are in severe pain should be treated with opioids as a first-line therapy, there's an increased need for complementary and alternative medicine (CAM) therapies during a time where opioids may not be readily prescribed to help manage pain.

A common CAM therapy used by adults with SCA is massage, which has been shown to be moderately effective for pain reduction. Another CAM therapy that has been shown to be moderately effective during pain crises is relaxation techniques to slow breathing and distract from the pain. In addition to other types of CAM, acupuncture can be used to stimulate the nervous system to relieve pain, nausea, neuropathy, anxiety, and depression. Acupuncture involves the use of

heat, needles, and pressure to certain points in the skin to cause physical responses in nerve cells. Acupuncture has been shown to help individuals with SCA, cancer, headaches, and back pain.

Implementation of a screening and care referral process for patients with SCA

Individuals with SCA tend to be at risk for complications, both physiologic and psychosocial, after being treated in a hospital, which can lead to difficulties with disease management. Care coordination refers to a long-term process of improving outcomes and organizing patient care activities for patients who need chronic care. For patients with SCA, the coordination team may consist of nurses, clinicians, and hematologists, and their care may occur in different environments.

Developing a specific screening process for individuals who come into the ED and are admitted to the hospital can help improve care and create more individualized treatment for patients with SCA. The screening process should assess the patient's care needs, most severe symptoms, pain management strategies, prescriptions, primary care and insurance information, and other common demographic information to help with individualized care planning and care transition management after leaving the ED.

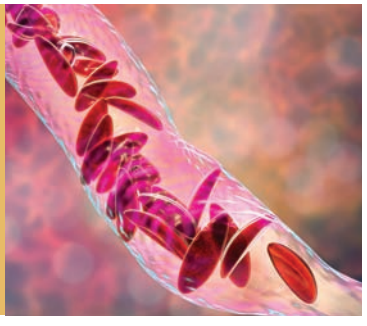
The use of opioids, I.V. fluids, and heat

Opioids continue to be the backbone of acute and chronic pain management for individuals with SCA. Morphine remains one of the most frequently used opioids for patients with SCA to manage vaso-occlusive crises. Morphine acts by increasing blood flow to areas that may have decreased blood flow. Despite the possible negative long-term effects of opioid use, morphine may be the most effective way to treat SCA pain. If the patient has an allergy to morphine, fentanyl can be used.

I.V. fluids are often used during treatment of patients with SCA. Patients who enter the hospital setting with vaso-occlusive pain episodes tend to be dehydrated. Although there's been some controversy over which type of I.V. fluid is optimal, studies have shown that the use of I.V. fluids can improve the hydration of deformed RBCs in patients with SCA.

Heat can be used to control pain stemming from vaso-occlusive crises. Pain is unique to every patient, but the use of heat may relax him or her and help lessen the pain. And heat is a method of pain management that can be used at home, increasing convenience for the patient.

Individuals with SCA tend to be at risk for complications, both physiologic and psychosocial, after being treated in a hospital, which can lead to difficulties with disease management.



Case study update

After completing his admission assessment and finding that Mr. E states a pain level of a 9/10, the nurse places a call to the hospitalist to request an alternate pain medication because Mr. E is experiencing a vaso-occlusive crisis that will require treatment with around-the-clock opioids. After conferring with the

on the web

CDC:
www.cdc.gov/ncbddd/sicklecell/facts.html

Mayo Clinic:
www.mayoclinic.org/diseases-conditions/sickle-cell-anemia/symptoms-causes/syc-20355876

MedlinePlus:
https://medlineplus.gov/sicklecelldisease.html

National Heart, Lung, and Blood Institute:
www.nhlbi.nih.gov/health-topics/sickle-cell-disease

US National Library of Medicine:
https://ghr.nlm.nih.gov/condition/sickle-cell-disease

hospitalist, morphine 2 mg I.V. every 2 hours around the clock is prescribed and the tramadol is discontinued.

For the next 24 hours, Mr. E rests and is treated with I.V. fluids, moist heat applied to his hands for 20 minutes every 4 hours, and morphine. By the second day, he's feeling much better, is afebrile, and only taking oral acetaminophen 650 mg every 6 hours as needed for pain.

Mr. E is discharged from the hospital on the third day with instructions about the importance of hydration, especially when playing sports and in the heat, along with a consult for a hematologist to better manage his SCA and consider the use of hydroxyurea therapy to minimize future vaso-occlusive crises. He's told to continue taking oral acetaminophen 650 mg every 6 hours as needed for pain, oral folate 400 mcg daily, and his previously prescribed home medications. In addition, the nurse advises Mr. E about the importance of meeting with a geneticist to discuss family planning in the future.

Patient-centered care

Care of patients with SCA experiencing vaso-occlusive crises has room for improvement related to patient outcomes and needs. Extreme pain compounded with potential bias and stigma creates a less-than-optimal healthcare experience for those in crisis. Nurses are in a unique position to help patients experiencing a sickle cell emergency by following best-practice recommendations and working to overcome barriers to care. ■

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INSTRUCTIONS

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