Managing sickle

Sickle cell disease is a chronic illness that impacts physical and social development, often requiring multiple hospitalizations. We give you the lowdown on treatments, collaborative care, and lifestyle adjustments to successfully manage this disease.

By Valerie Gill, MSN • Assistant Professor • Kingsborough Community College • Brooklyn, N.Y.

Joanne Lavin, EdD • Professor and Director of Nursing Programs • York College • Jamaica, N.Y.

Maria Sim, EdD • Associate Professor and Director of RN-BSN Program • State University of New York Downstate Medical Center College of Nursing • Brooklyn, N.Y.

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Michelle, a 14-year-old Black American, is admitted to your unit with severe abdominal pain. She was diagnosed with sickle cell disease (SCD) as an infant. Her vital signs are: temperature, 100.4° F (38° C); heart rate, 100 beats/minute; respiratory rate, 30 breaths/minute; oxygen saturation, 90%; and BP, 95/72 mm Hg. She's slightly underweight for her age and has swollen feet. A complete blood cell (CBC) count reveals: hemoglobin, 8.0 g/dL; hematocrit, 24%; white blood cell count, 12,000 cells/mm^3; and platelet count, 140,000 cells/mm^3. Michelle has experienced several sickle cell crises over the years. However, they've become more frequent since she became an adolescent. Michelle and her parents are very knowledgeable about SCD and the immediate and long-term care required to manage the challenges of this condition.

SCD is an inherited disorder in which there are inadequate healthy red blood cells (RBCs) to transport oxygen to the body's tissues. Normally RBCs are flexible and round, traveling easily through blood vessels. In SCD, the RBCs are shaped like a half moon (or sickle) and rigid, obstructing capillary blood flow and causing severe pain (see *Picturing SCD*). Microscopic obstruction leads to engorgement, tissue ischemia, hypoxia, and ultimately large infarctions. Repeated sickle cell crises slowly destroy organs and tissues. The spleen and kidneys are especially prone to organ damage.

In this article, we'll discuss treatments, collaborative care, and lifestyle modifications to successfully manage SCD. But first, let's take a closer look at how SCD develops and its signs and symptoms.

A matter of inheritance

SCD results from a mutation in the gene responsible for hemoglobin production. Under normal circumstances healthy hemoglobin A is produced; people who have SCD produce sickle hemoglobin S. The sickle gene is transmitted in families via autosomal-recessive inheritance. This mutated gene is common among people with African, South American, Mediterranean, Middle Eastern, and East Indian ancestry. In the United States, it occurs most often in

cell disease

Black American and Hispanic individuals. According to the National Heart, Lung, and Blood Institute, SCD affects an estimated 70,000 to 100,000 people in the United States: about 1 of every 500 Black American and 1 of every 36,000 Hispanic births.

There are three main types of SCD:

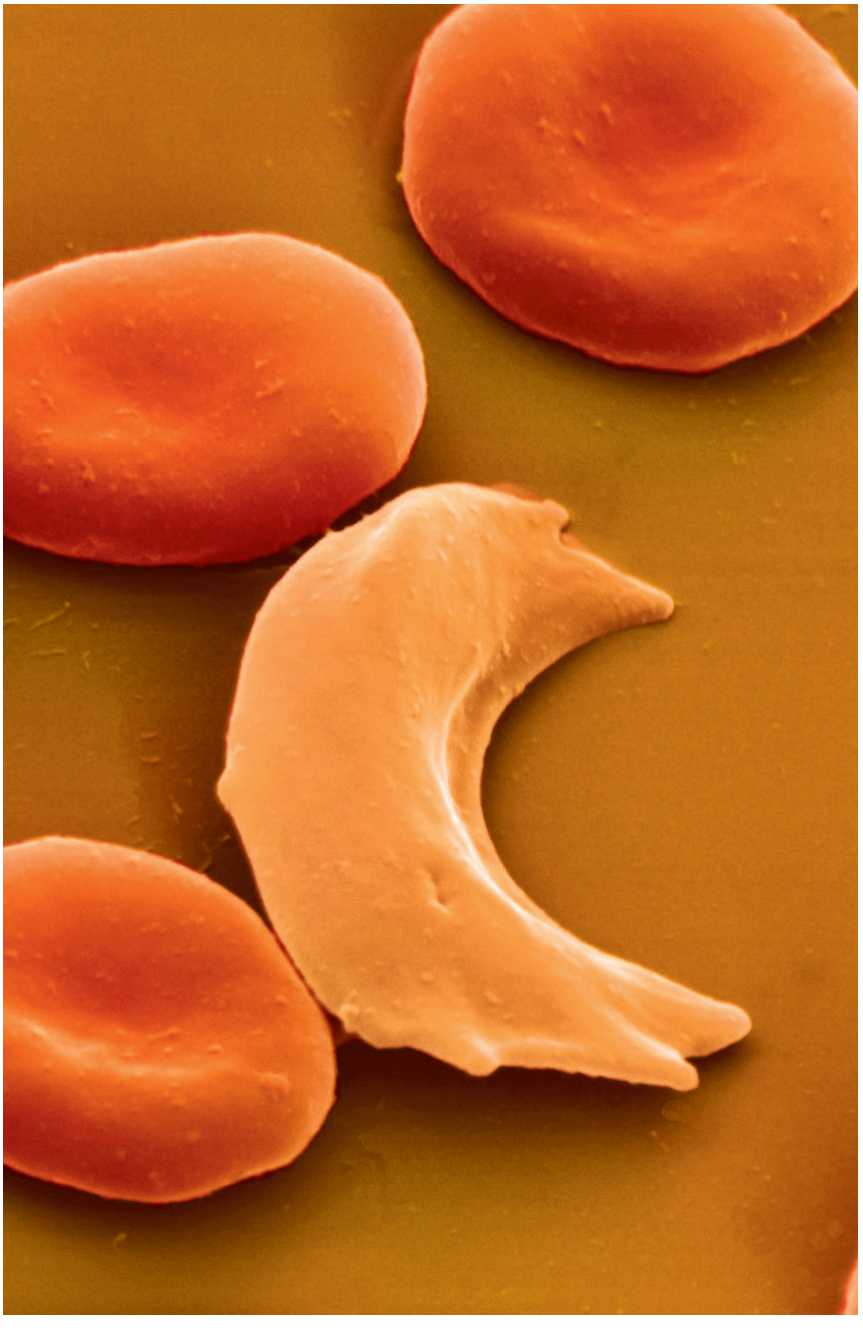
• ***hemoglobin SS disease.*** Individuals have two copies of the hemoglobin S gene. This is the most common type of SCD (65%) and results in an average life span of 45 years.

• ***hemoglobin SC disease.*** Individuals have one copy of the hemoglobin S gene and one copy of the hemoglobin A gene. The anemia is generally milder because the RBCs are C-shaped. This is the second most common type of SCD (25%) and results in an average life span of 65 years.

• ***hemoglobin SB (beta) thalassemia disease including SB0 (beta zero) and SB+ thalassemia disease.*** Individuals have one copy of the hemoglobin S gene and one copy of the hemoglobin beta-thalassemia gene. The beta-thalassemia genes results in reduced (B+) or lack of expression (B0) of normal hemoglobin A. Thus, the person has either only hemoglobin S or mostly hemoglobin S with a small percentage of normal hemoglobin A. The typical life span is in the mid-50s.

People with sickle cell trait have one normal hemoglobin gene and one defective sickle gene. These individuals typically don't experience symptoms because the normal gene results in some production of normal hemoglobin. About 2 million Americans have sickle cell trait, which occurs in 1 in 12 Black Americans, according to the National Heart, Lung, and Blood Institute.

If two people with sickle cell trait have a child there will be:



• a 25% chance of an unaffected child with normal hemoglobin
• a 50% chance of a child who, like the parents, has one normal and one sickle gene (a carrier of sickle cell trait)
• a 25% chance of a child who has SCD because of two inherited sickle genes.

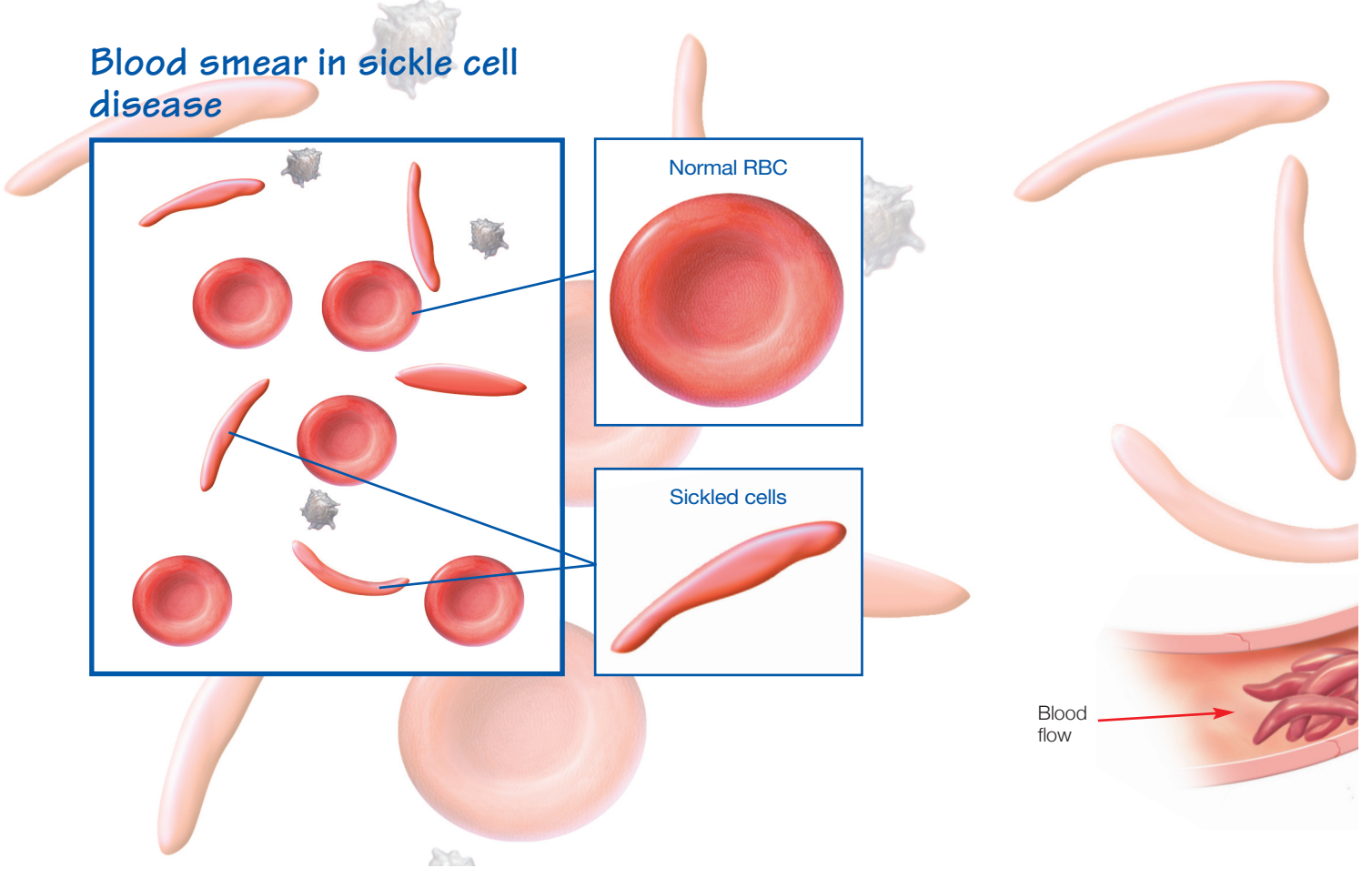
If a person with SCD has a child with a person who has normal hemoglobin genes, their child will have sickle cell trait. If two people with SCD have a child, the child will have SCD.

A lifetime of symptoms

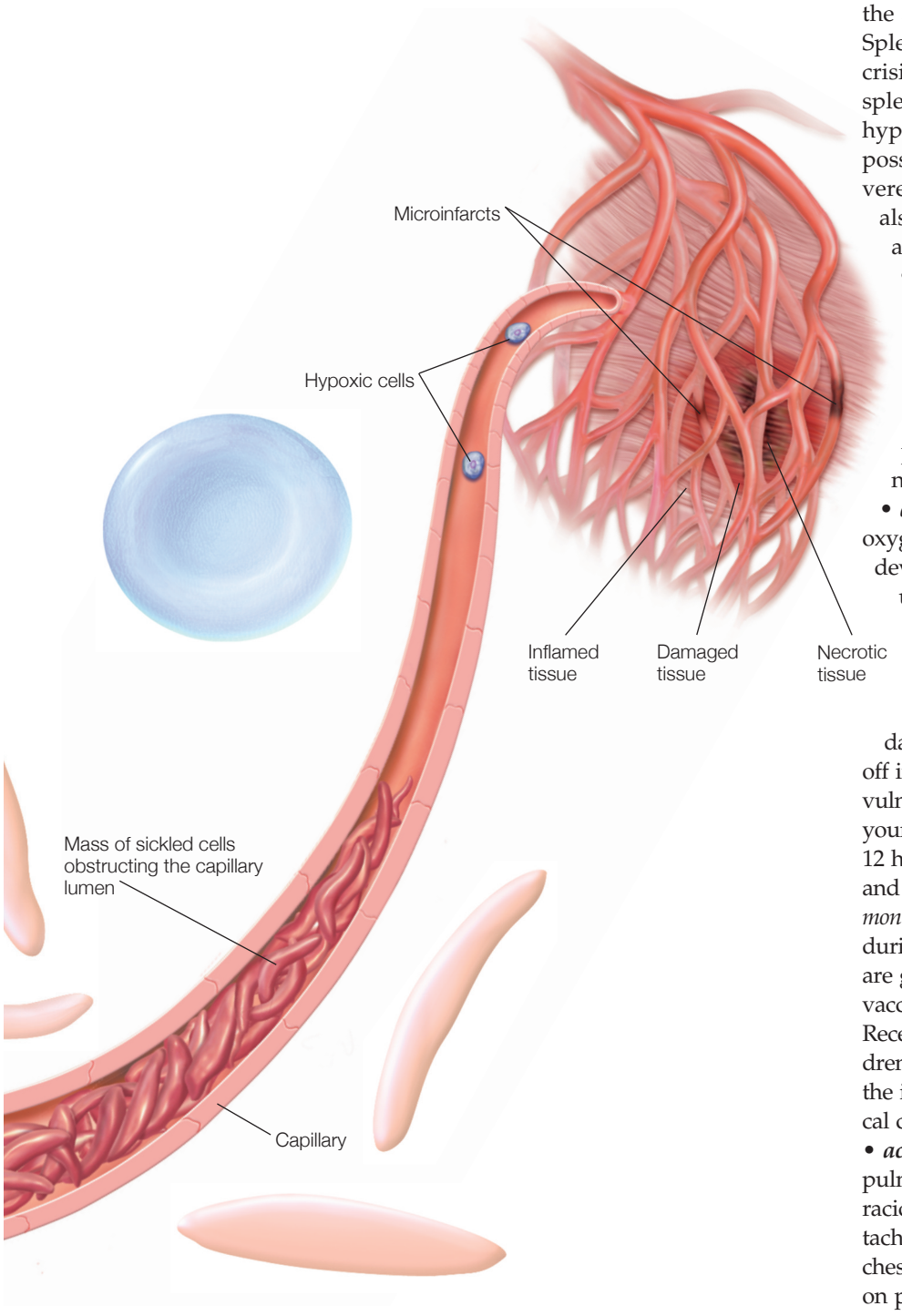
SCD usually manifests after 4 to 6 months of age because initial sickling is inhibited by high levels of fetal hemoglobin. Typical signs and symptoms for individuals with SCD include (see *Clinical manifestations of SCD*):

• ***pain episodes.*** Pain occurs when the irregularly shaped RBCs cause stasis of the blood with clumping of cells in the microcirculation, ischemia, and infarction. This vaso-occlusive crisis is the most

Picturing SCD



Sickle cell crisis



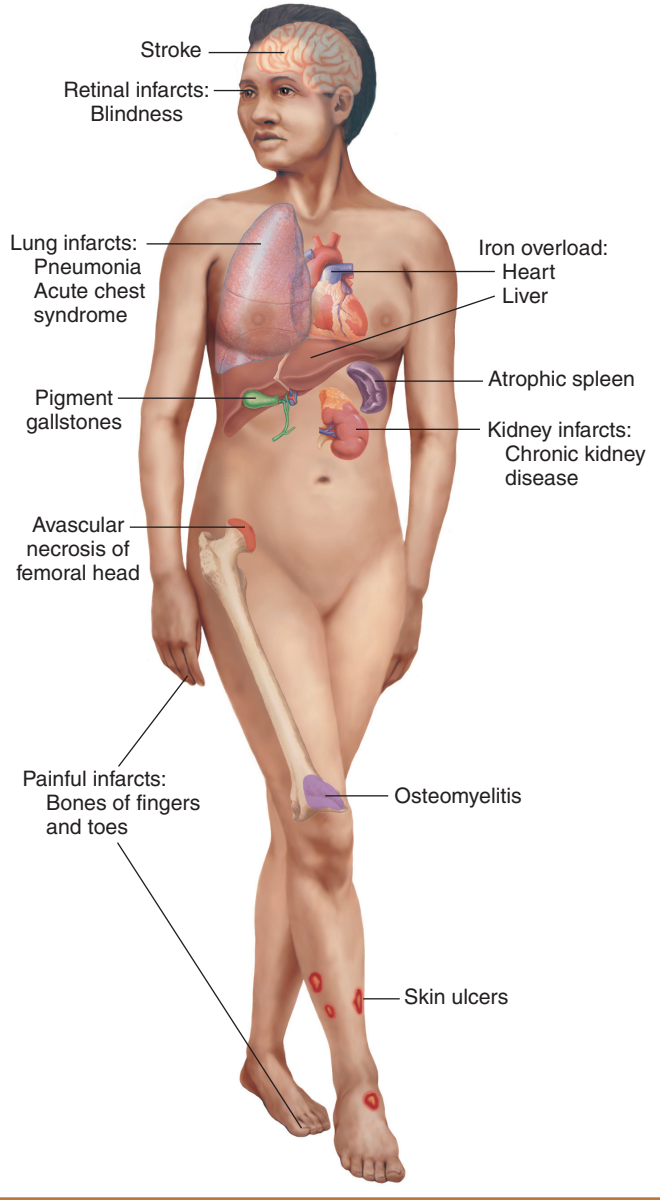
common type of sickle cell crisis and lasts for days or weeks. It frequently occurs in the abdomen, back, joints, and chest areas. Splenic sequestration is a life-threatening crisis, causing pooling of blood in the spleen and resulting in profound anemia, hypovolemia, cardiovascular collapse, and possible death. Pain episodes are often severe and require hospitalization. Patients also experience psychological symptoms associated with chronic, recurring pain.

- ***anemia.*** Sickle cells are fragile, breaking apart easily and dying after 10 to 20 days. Normal RBCs live for about 120 days. This results in a decreased amount of oxygen delivery to tissues. Individuals typically experience fatigue, pallor, tachypnea, jaundice, and shortness of breath when anemia is severe.
- ***delayed growth.*** RBCs provide needed oxygen and nutrients for growth and development. Children with SCD are often underweight and have delays in development, including puberty. Frequent hospitalizations may interfere with school and social development.
- ***frequent infections.*** Sickle cells can damage the spleen, an organ that fights off infections. Children with SCD are more vulnerable to bacterial infections. Infants and young children can actually die in as little as 12 hours from the onset of fever. Septicemia and meningitis caused by *Streptococcus pneumoniae* are the most common causes of death during early childhood. Infants and children are given antibiotics and the pneumococcal vaccine to prevent life-threatening infections. Recent decreases in mortality in Black children under age 4 with SCD coincided with the introduction of the 7-valent pneumococcal conjugate vaccine in 2000.
- ***acute chest syndrome.*** This describes new pulmonary findings on X-ray such as thoracic cage infarction of bones. Signs include tachypnea and shallow respirations. The chest may have slight swelling and warmth on palpation; erythema may indicate rib or vertebral infarction. The individual

may also report pain, cough, fever, and abdominal pain.

• ***hand-foot syndrome.*** An early indication of SCD is swollen hands and feet. This occurs because the sickle shaped cells prevent blood flow out of these extremities. Pain, mobility, and activity impairments result.

Clinical manifestations of SCD



• ***vision problems.*** Problems with vision occur if the sickle shaped cells block the flow of blood in the small vessels in the eyes. These cells can actually damage the retina, uvea, and optic nerve. Proliferative sickle cell retinopathy may cause vitreous hemorrhage and retinal detachment with loss of vision.

• ***hepatomegaly.*** Liver involvement is present in 40% to 80% of individuals with SCD. Acute attacks with pain in the right upper abdominal quadrant and hemolysis lead to the production of gallstones.

• ***stroke.*** Stasis and clumping of the irregularly shaped RBCs can result in cerebral occlusion and stroke manifested by headache, vomiting, seizure, weakness, paralysis, and motor disturbances.

• ***priapism.*** Men of any age with SCD may experience painful erection of the penis that can last for hours, days, or weeks. This usually occurs during sleep and is often associated with dehydration, hypoventilation, or a full bladder. Men with SCD are also at increased risk for erectile dysfunction.

• ***psychosocial issues.*** Depression and disturbed self-concept associated with a chronic disease often occur in individuals with SCD.

Diagnosis and screening

Hemoglobin electrophoresis of cord blood can be used to screen newborns for SCD. The sickle turbidity test—a finger-stick test with a rapid result—is used to screen children ages 6 and up. Hemoglobin electrophoresis can then confirm the diagnosis. DNA analysis provides the most accurate diagnosis, but it's expensive and not widely available.

Routine hematologic tests (CBC and ferritin levels) are done to evaluate anemia. The child's height and weight are routinely measured to identify failure to thrive. Routine assessment includes a history of pain and signs of infection and inflammation. As the child becomes older, genetic counseling will be needed.

Supportive and preventive treatments

Treatment focuses on managing symptoms and preventing crises and complications. Standard medications used to treat SCD include:

• ***analgesics.*** Patients regularly take over-the-counter (OTC) pain-relieving medications such as ibuprofen; during a crisis, stronger opioid analgesics, such as morphine or hydromorphone, are prescribed. In addition to opioids, ketorolac is effective in reducing pain stimulus and inflammation.

• ***antibiotics.*** Children with SCD are placed on antibiotics as infants to prevent pneumonia.

• ***hydroxyurea.*** A cell-cycle-phase antineoplastic drug that inhibits DNA synthesis, this medication reduces the frequency of painful crises. However, it can produce bone marrow suppression and worsen anemia.

Nonpharmacologic measures include heat applications and limiting movement of the painful extremity. It's essential that the child be included in the pain management plan to ensure a sense of control and prevent psychological trauma associated with pain syndromes.

Blood transfusions are often needed to treat anemia. A sudden worsening of anemia because of infection or spleen enlargement is an indicator for a blood transfusion. Blood transfusions improve tissue oxygenation and reduce sickling; however, frequent blood transfusions can lead to iron toxicity.

Experimental treatments include:

• ***gene therapy.*** Researchers are exploring the possibility of inserting a normal gene into the bone marrow of children with SCD to promote the production of normal hemoglobin.

• ***butyric acid.*** This food additive increases the amount of fetal hemoglobin in the blood.

• ***clotrimazole.*** This OTC medication used to treat fungal infections helps prevent a loss of water from RBCs and may reduce the formation of sickle cells.

Complications of SCD

cheat sheet

- Frequent infections
- Stroke
- Pneumonia
- Acute chest syndrome and thoracic cage infarction
- Osteomyelitis
- Avascular necrosis
- Heart disease
- Chronic renal failure
- Septicemia
- Meningitis
- Sickle cell retinopathy
- Gallstones and hepatomegaly
- Chronic leg ulcers

• ***nitric oxide.*** Individuals with SCD have decreased levels of nitric oxide. This gas helps keep blood vessels open and reduces the stickiness of RBCs. It may also reduce the clumping of sickle cells during crises.

• ***nicosan.*** This herb is used in Nigeria to prevent episodes of sickle cell crisis.

• ***decitidine.*** This medication increases hemoglobin F levels, a type of hemoglobin that carries more oxygen.

• ***GMI-1070.*** An experimental pan-selectin inhibitor that treats vaso-occlusive crises, this medication inhibits a key early step in the inflammatory processes involved with cell adhesion and restores blood flow to tissues.

• ***bone marrow transplant.*** Bone marrow used for transplant must come from a matched donor—usually a family member who doesn't have SCD. Bone marrow transplant is a potentially curative treatment.

Strategize for success

Nursing strategies for SCD include:

• ***Manage acute crises.*** During an acute crisis, comfort measures, use of analgesics, and complementary approaches such as massage and distraction are key. Healthcare providers focus on hydration, prevention of

infections, and early recognition of complications. Pharmacologic treatment is started immediately to manage pain. Some individuals need blood transfusions to manage severe anemia. Indications for blood transfusions include splenic sequestration, acute chest syndrome, severe anemia, and stroke.

- ***Teach effective pain management measures.*** Pain is a recurring factor in SCD. Pharmacologic and nonpharmacologic measures to control and/or decrease the severity of pain episodes are essential. These include physical and occupational therapy, physiotherapy, cognitive-behavioral therapy, and support groups. If your patient is a child, her parents need to understand effective measures and provide them to control pain. Reading and watching TV/videos can be effective distraction techniques. For the teen exploring hobbies, choosing physical activities such as swimming, especially in a heated pool, can improve pain management and physical stamina while limiting stress on joints.
- ***Teach effective lifestyle modifications.*** It's crucial that children and adults who have SCD learn effective lifestyle modifications. Parents need to encourage their child to engage in normal childhood activities when possible. Play is crucial, but some activities may need to be modified. For the teen, the task is to recognize what triggers a crisis, manage or eliminate these factors, and compensate for less appropriate activities with alternatives that bring a sense of accomplishment and enjoyment (see *Treating triggers*).
- ***Instruct when to contact the healthcare provider.*** Symptoms to be reported include fever; swelling of the hands or feet; swelling of the abdomen and lower left side with tenderness; pain in the joints, chest, or muscles; pale skin or nail beds or a yellowish skin color (jaundice); sudden fatigue with less interest in surroundings; and an erection of the penis that won't go away.
- ***Teach stress management techniques.*** Stress triggers sickle cell crises and decreases coping abilities. Learning how to effectively reduce stress through breathing, muscle relaxation, meditation, and guided imagery can assist your patient in gaining a sense of control. Maintaining a balance of nutrition, rest, and exercise is also important. A child with SCD may not yet be able to identify how to effectively balance rest and exercise, so parents need to manage this. A teen with SCD should be encouraged to participate in the process. Joining a support group for people who have SCD or chronic diseases may be helpful. The use of social networking sites such as Facebook and Twitter may decrease isolation and stress.
- ***Encourage the acknowledgment of feelings.*** Many people living with SCD find it helpful to line up supportive individuals, such as parents, teachers, and friends. Others talk with a therapist or join a support group specifically for people with their condition. It's important for your patient to acknowledge her emotions, talk about feelings of anger and dependence, and recognize these as part of the illness. Encourage strategies that increase a sense of control over your patient's life to decrease feelings of powerlessness and hopelessness. Families may become overprotective of their children, stifling their growth and ability to manage their disease. Adolescents, especially, need to be allowed to determine treatment strategies. Teens may be concerned with, and need to discuss, body image issues related to delayed sexual maturity.

Cognitive-behavioral therapies teach coping skills and result in less negative thinking and even less pain. Cognitive therapy helps a person identify thoughts, beliefs, and perceptions about a situation, as well as how these impact behaviors. For example, pain can be perceived negatively as a sign of a problem or positively as an indication that one's body recognizes the need to do something different. The goal is to help patients

Treating triggers

Trigger	Considerations
Infection	Obtain pneumonia and annual flu vaccinations.
Dehydration	Drink at least eight glasses of water daily to keep blood diluted and reduce sickling.
Excessive heat/cold	Avoid temperature extremes because of resultant vasoconstriction.
Nutritional deficiencies	Eat a balanced diet and take vitamin supplements. Adequate nutrition and vitamins, especially folic acid, help make new RBCs.
Decreased oxygenation	Avoid high-altitude areas and unpressurized airplanes, which lower oxygen levels.
Certain medications	Avoid decongestants, such as pseudoephedrine, which can tighten blood vessels and prevent the smooth flow of RBCs through the vessels.
Excessive exercise or physical activity	Determine appropriate exercise levels and activities.

adapt and learn to manage their illness rather than allowing the disease to control them.

Let's return to our patient, Michelle. Initial priorities include pain management and fluids.

- For the first 2 days, Michelle receives patient-controlled analgesia. You monitor for pain relief and carefully position her, as well as determine what pain measures worked in the past.
- Hydration is promoted through oral and I.V. therapy. You monitor intake and output, weight, and serum electrolytes. Additionally, you encourage nutrition, providing Michelle with favorite foods and frequent snacks because she's underweight. Vitamin supplementation (folic acid) is also prescribed.
- Energy expenditure is minimized through rest because of Michelle's decreased hemoglobin and oxygen saturation levels. You administer short-term oxygen therapy and monitor her respiratory function.
- Blood replacement therapy (transfusion) is provided to treat the anemia and reduce the viscosity of the sickled blood cells.
- You administer prescribed antibiotics to treat Michelle's infection.
- You provide emotional support to Michelle and her parents and reinforce their knowledge of the disease and prevention of crises.

Surviving with SCD

SCD is a hereditary hemoglobinopathy characterized by abnormal hemoglobin cells. These sickle cells cause occlusion of small blood vessels, ischemia, severe pain episodes, delayed growth and development, and damage to many organs. SCD requires a multidisciplinary approach to manage painful crisis episodes, prevent complications, decrease emotional problems, and promote a healthy lifestyle. Early screening and intervention with prophylactic antibiotics and intensive patient and family education can increase life expectancy and quality. ■

Learn more about it

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On the web

These online resources may be helpful to your patients and their families:

KidsHealth.org: http://kidshealth.org/parent/medical/heart/sickle_cell_anemia.html
MedlinePlus: http://www.nlm.nih.gov/medlineplus/sicklecellanemia.html
National Heart, Lung, and Blood Institute: http://www.nhlbi.nih.gov/health/dci/Diseases/Sca/SCA_WhatIs.html
Sickle Cell Disease Association of America: http://www.sicklecelldisease.org
U.S. National Library of Medicine Genetics Home Reference: http://ghr.nlm.nih.gov/condition/sickle-cell-disease.

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Managing sickle cell disease

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