

SICKLE CELL CRISIS

Sickle cell disease (sickle hemoglobinopathies) is a group of genetic diseases, the most common forms being homozygous hemoglobin SS disease (sickle cell anemia), hemoglobin SC disease, and sickle [beta]-thalassemia.

Sickle cell disease primarily affects black populations and people of Mediterranean, South/Central American, Arabian, and East Indian descent. It renders the individual vulnerable to repeated painful crises that can progressively destroy vital organs. These crises are:

Vaso-occlusive/thrombotic crisis: Related to infection, dehydration, fever, hypoxia, and characterized by multiple infarcts of bones, joints, and other target organs, with tissue pain and necrosis caused by plugs of sickled cells in the microcirculation.

Hypoplastic/aplastic crisis: May be secondary to severe (usually viral) infection or folic acid deficiency, resulting in cessation of production of RBCs and bone marrow.

Hyperhemolytic crisis: Reticulocytes are increased in peripheral blood; and bone marrow is hyperplastic. Characterized by anemia and jaundice (effects of hemolysis).

Sequestration crisis (more commonly occurs in infants): Massive, sudden erythrocytosis with pooling of blood in the viscera (splenomegaly), resulting in hypovolemic shock/possible death. This crisis occurs in patients with intact splenic function.

CARE SETTING

Sickle cell disease is generally managed at the community level, with many of the interventions included here being appropriate for this focus; however, this plan of care addresses sickle cell crisis, which usually requires hospitalization during the acute phase.

RELATED CONCERNS

Cerebrovascular accident (CVA)/stroke

Cholecystitis with cholelithiasis

Chronic obstructive pulmonary disease (COPD) and asthma

Cirrhosis of the liver

Heart failure: chronic

Pneumonia: microbial

Psychosocial aspects of care

Seizure disorders

Sepsis/septicemia

Patient Assessment Database

Depends on severity of condition, presence of complications.

ACTIVITY/REST

- May report:** Lethargy, fatigue, weakness, general malaise
Loss of productivity; decreased exercise tolerance; greater need for sleep and rest
- May exhibit:** Listlessness; severe weakness and increasing pallor (aplastic crisis)
Gait disturbances (pain, kyphosis, lordosis); inability to walk (pain)
Poor body posture (slumping of shoulders indicative of fatigue)
Decreased range of motion (ROM) (swollen, inflamed joints); joint, bone deformities
Generalized retarded growth; tower-shaped skull with frontal bossing; disproportionately long arms and legs; short trunk; narrowed shoulders/hips; and long, tapered fingers

CIRCULATION

- May report:** Palpitations or anginal chest pain (concomitant coronary artery disease [CAD]/myocardial ischemia, acute chest syndrome)
Intermittent claudication
- May exhibit:** Apical pulse: Point of maximal impulse (PMI) may be displaced to the left (cardiomegaly)
Tachycardia, dysrhythmias (hypoxia), systolic murmurs

BP: Widened pulse pressure
Generalized symptoms of shock, e.g., hypotension, rapid thready pulse, and shallow respirations (sequestration crisis)
Peripheral pulses throbbing on palpation
Bruit (reflects compensatory mechanisms of anemia; may also be auscultated over the spleen because of multiple splenic infarcts)
Capillary refill delayed (anemia or hypovolemia)
Skin color: Pallor or cyanosis of skin, mucous membranes, and conjunctiva. (*Note:* Pallor may appear as yellowish brown color in brown-skinned patients and as ashen gray in black-skinned patients). Jaundice: Scleral icterus, generalized icteric coloring (excessive RBC hemolysis)
Dry skin/mucous membranes
Diaphoresis (either sequestration or vaso-occlusive crisis; acute pain or shock)

ELIMINATION

May report: Frequent voiding, voiding in large amounts, nocturia
May exhibit: Right upper quadrant (RUQ) abdominal tenderness, enlargement/distension (hepatomegaly); ascites
Left upper quadrant (LUQ) abdominal fullness (spleen may be enlarged or may be atrophic and nonfunctional from repeated splenic infarcts and fibrosis)
Dilute, pale, straw-colored urine; hematuria or smoky appearance (multiple renal infarcts)
Urine specific gravity decreased (may be fixed with progressive renal disease)

EGO INTEGRITY

May report: Resentment and frustration with disease, fear of rejection from others
Negative feelings about self, ability to deal with life/situation.
Concern regarding being a burden to significant others (SOs); financial concerns, possible loss of insurance/benefits, lost time at work/school; fear of genetic transmission of disease
May exhibit: Anxiety, restlessness, irritability, apprehension, withdrawal, narrowed focus, self-focusing, unresponsiveness to questions, regression, depression, decreased self-concept
Dependent relationship with whoever can offer security and protection

FOOD/FLUID

May report: Thirst
Anorexia, nausea/vomiting
May exhibit: Height/weight usually in the lower percentiles
Poor skin turgor with visible tenting (crisis, infection, and dehydration)
Dry skin/mucous membranes
Jugular venous distension (JVD) and general peripheral edema (concomitant HF)

HYGIENE

May report: Difficulty maintaining ADLs (pain or severe anemia)
May exhibit: Unkempt appearance, poor personal hygiene

NEUROSENSORY

May report: Headaches or dizziness
Visual disturbances (e.g., hemianopsia, retinopathy, nystagmus)
Tingling in the extremities
Disturbances in pain and position sense
May exhibit: Mental status usually unaffected except in cases of severe sickling (cerebral infarction/intracranial hemorrhage)
Weakness of the mouth, tongue, and facial muscles; aphasia (in cerebral infarction of dominant hemisphere)
Abnormal reflexes, decreased muscle strength/tone; abnormal involuntary movements; hemiplegia or sudden hemiparesis, quadriplegia
Ataxia, seizures

Meningeal irritation (intracranial hemorrhage), e.g., decreasing level of consciousness (LOC), nuchal rigidity, focal neurological deficits, vomiting, severe headache

PAIN/DISCOMFORT

- May report:** Pain as severe, throbbing, gnawing of varied location (localized, migratory, or generalized)
Recurrent, sharp, transient headaches
Back pain (changes in vertebral column from recurrent infarctions); joint/bone pain accompanied by warmth, tenderness, erythema, and occasional effusions (vaso-occlusive crisis); deep bone infarctions may have no apparent signs of irritation
Gallbladder tenderness and pain (excessive accumulation of bilirubin as a result of increased erythrocyte destruction)
- May exhibit:** Sensitivity to palpation over affected areas
Guarding/holding joints in position of comfort; decreased ROM (result of joint pain and swelling)
Maladaptive pain behaviors, e.g., guilt for being ill, denial of any aspect of disease, indulgence in precipitating factors (overwork, strenuous exercise)

RESPIRATION

- May report:** Dyspnea on exertion or at rest
History of repeated pulmonary infections/infarctions, pulmonary fibrosis, pulmonary hypertension or cor pulmonale
- May exhibit:** Acute respiratory distress, e.g., dyspnea, chest pain, and cyanosis (especially in crisis)
Bronchial/bronchovesicular sounds in lung periphery; diminished breath sounds (pulmonary fibrosis)
Crackles, rhonchi, wheezes, diminished breath sounds (HF)
Increased anteroposterior (AP) diameter of the chest (barrel chest)

SAFETY

- May report:** History of repeated/frequent transfusions
- May exhibit:** Low-grade fever
Impaired vision (sickle retinopathy), decreased visual acuity (temporary/permanent blindness)
Leg ulcers (common in adult patients, especially found on the internal and external malleoli and the medial aspect of the tibia)
Lymphadenopathy

SEXUALITY

- May report:** Loss of libido; amenorrhea; priapism, impotence
- May exhibit:** Delayed sexual maturity
Pale cervix and vaginal walls (anemia)

TEACHING/LEARNING

- May report:** History of HF (chronic anemic state); pulmonary hypertension or cor pulmonale (multiple pulmonary infections/infarctions); chronic leg ulcers, delayed healing
- Discharge plan considerations:** **DRG projected mean length of inpatient stay: 4.3 days**
May need assistance with shopping, transportation, self-care, homemaker/maintenance tasks
Refer to section at end of plan for postdischarge considerations.

DIAGNOSTIC STUDIES

CBC: Reticulocytosis (count may vary from 30%–50%); leukocytosis (especially in vaso-occlusive crisis), with counts over 20,000 indicate infection, decreased Hb (5–10 g/dL) and total RBCs, elevated platelets, and a normal to elevated MCV.

Stained RBC examination: Demonstrates partially or completely sickled, crescent-shaped cells; anisocytosis; poikilocytosis; polychromasia; target cells; Howell-Jolly bodies; basophilic stippling; occasional nucleated RBCs (normoblasts).

Sickle-turbidity tube test (Sicklelex): Routine screening test that determines the presence of hemoglobin S (HbS) but does not differentiate between sickle cell anemia and trait.

Hemoglobin electrophoresis: Identifies any abnormal hemoglobin types and differentiates between sickle cell trait and sickle cell anemia. Results may be inaccurate if patient has received a blood transfusion within 3–4 mo before testing.

ESR: Elevated.

Erythrocyte fragility: Decreased (osmotic fragility or RBC fragility); RBC survival time decreased (accelerated breakdown).

ABGs: May reflect decreased PO₂ (defects in gas exchange at the alveolar capillary level); acidosis (hypoxemia and acidic states in vaso-occlusive crisis).

Serum bilirubin (total and indirect): Elevated (increased RBC hemolysis).

Acid phosphatase (ACP): Elevated (release of erythrocytic ACP into the serum).

Alkaline phosphatase: Elevated during vaso-occlusive crisis (bone and liver damage).

LDH: Elevated (RBC hemolysis)

Serum potassium and uric acid: Elevated during vaso-occlusive crisis (RBC hemolysis).

Serum iron: May be elevated or normal (increased iron absorption due to excessive RBC destruction).

Total iron-binding capacity (TIBC): Normal or decreased.

Urine/fecal urobilinogen: Increased (more sensitive indicators of RBC destruction than serum levels).

Intravenous pyelogram (IVP): May be done to evaluate kidney damage.

Bone radiographs: May demonstrate skeletal changes, e.g., osteoporosis, osteosclerosis, osteomyelitis, or avascular necrosis.

X-rays: May indicate bone thinning, osteoporosis.

NURSING PRIORITIES

1. Promote adequate cellular oxygenation/perfusion.
2. Alleviate pain.
3. Prevent complications.
4. Provide information about disease process/prognosis, and treatment needs.

DISCHARGE GOALS

1. Oxygenation/perfusion adequate to meet cellular needs.
2. Pain relieved/controlled.
3. Complications prevented/minimized.
4. Disease process, future expectations, potential complications, and therapeutic regimen understood.
5. Plan in place to meet needs after discharge.

NURSING DIAGNOSIS: Gas Exchange, impaired

May be related to

Decreased oxygen-carrying capacity of the blood, reduced RBC life span/premature destruction, abnormal RBC structure; sensitivity to low oxygen tension (strenuous exercise, increase in altitude)

Increased blood viscosity (occlusions created by sickled cells packing together within the capillaries) and pulmonary congestion (impairment of surface phagocytosis)

Predisposition to bacterial pneumonia, pulmonary infarcts

Possibly evidenced by

Dyspnea, use of accessory muscles

Restlessness, confusion

Tachycardia

Cyanosis (hypoxia)

DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:

Respiratory Status: Gas Exchange (NOC)

Demonstrate improved ventilation/oxygenation as evidenced by respiratory rate within normal limits, absence of cyanosis and use of accessory muscles; clear breath sounds.

Participate in ADLs without weakness and fatigue.

Display improved/normal pulmonary function tests.

ACTIONS/INTERVENTIONS	RATIONALE
<p>Respiratory Monitoring (NIC)</p>	
<p>Independent</p>	
<p>Monitor respiratory rate/depth, use of accessory muscles, areas of cyanosis.</p>	<p>Indicators of adequacy of respiratory function or degree of compromise and therapy needs/effectiveness.</p>
<p>Auscultate breath sounds, noting presence/absence, and adventitious sounds.</p>	<p>Development of atelectasis and stasis of secretions can impair gas exchange.</p>
<p>Monitor vital signs; note changes in cardiac rhythm.</p>	<p>Compensatory changes in vital signs and development of dysrhythmias reflect effects of hypoxia on cardiovascular system.</p>
<p>Investigate reports of chest pain and increasing fatigue. Observe for signs of increased fever, cough, adventitious breath sounds.</p>	<p>Reflective of developing acute chest syndrome (i.e., chest pain, dyspnea, fever, and leukocytosis), which increases the workload of the heart and oxygen demand.</p>
<p>Assess level of consciousness/mentation regularly.</p>	<p>Brain tissue is very sensitive to decreases in oxygen and may be an early indicator of developing hypoxia.</p>
<p>Ventilation Assistance (NIC)</p>	
<p>Assist in turning, coughing, and deep-breathing exercises.</p>	<p>Promotes optimal chest expansion, mobilization of secretions, and aeration of all lung fields; reduces risk of stasis of secretions/pneumonia.</p>
<p>Evaluate activity tolerance; limit activities to those within patient tolerance or place patient on bedrest. Assist with ADLs and mobility as needed.</p>	<p>Reduction of the metabolic requirements of the body reduces the oxygen requirements/degree of hypoxia.</p>
<p>Encourage patient to alternate periods of rest and activity. Schedule rest periods as indicated.</p>	<p>Protects from excessive fatigue, reduces oxygen demands/degree of hypoxia.</p>
<p>Demonstrate and encourage use of relaxation techniques, e.g., guided imagery and visualization.</p>	<p>Relaxation decreases muscle tension and anxiety and hence the metabolic demand for oxygen.</p>
<p>Promote adequate fluid intake, e.g., 2–3 L/day within cardiac tolerance.</p>	<p>Sufficient intake is necessary to provide for mobilization of secretions and to prevent hyperviscosity of blood/capillary occlusion.</p>
<p>Screen health status of visitors/staff.</p>	<p>Protects from potential sources of respiratory infection.</p>
<p>Collaborative</p>	
<p>Administer supplemental humidified oxygen as indicated.</p>	<p>Maximizes oxygen transport to tissues, particularly in presence of pulmonary insults/pneumonia. <i>Note:</i> Oxygen should be given only in the presence of confirmed hypoxemia because oxygen can suppress erythropoietin levels, further reducing the production of RBCs.</p>

ACTIONS/INTERVENTIONS	RATIONALE
<p>Respiratory Monitoring (NIC)</p> <p>Collaborative</p> <p>Monitor laboratory studies, e.g., CBC, cultures, ABGs/pulse oximetry, chest x-ray, pulmonary function tests.</p> <p>Perform/assist with chest physiotherapy, intermittent positive-pressure breathing (IPPB), and incentive spirometer.</p> <p>Administer packed RBCs (PRCs) or exchange transfusions as indicated.</p> <p>Administer medications as indicated: Antipyretics, e.g., acetaminophen (Tylenol):</p> <p>Antibiotics.</p>	<p>Patients are particularly prone to pneumonia, which is potentially fatal because of its hypoxemic effect of increasing sickling.</p> <p>Mobilizes secretions and increases aeration of lung fields.</p> <p>Increases number of oxygen-carrying cells, dilutes the percentage of HbS to prevent sickling, improves circulation, and removes/decreases number of sickled cells. PRCs are usually used because they are less likely to create circulatory overload. <i>Note:</i> Partial transfusions are sometimes used prophylactically in high-risk situations, e.g., chronic, severe leg ulcers, preparation for general anesthesia, third trimester of pregnancy.</p> <p>Maintains normothermia to reduce metabolic oxygen demands without affecting serum pH, which may occur with aspirin.</p> <p>A broad-spectrum antibiotic is started immediately pending culture results of suspected infections, then may be changed when the specific pathogen is identified.</p>

<p>NURSING DIAGNOSIS: Tissue Perfusion, ineffective (specify)</p> <p>May be related to</p> <p>Vaso-occlusive nature of sickling, inflammatory response Arteriovenous (AV) shunts in both pulmonary and peripheral circulation Myocardial damage from small infarcts, iron deposits, and fibrosis</p> <p>Possibly evidenced by</p> <p>Changes in vital signs; diminished peripheral pulses/capillary refill; general pallor Decreased mentation, restlessness Angina, palpitations Tingling in extremities, intermittent claudication, bone pain Transient visual disturbances Ulcerations of lower extremities, delayed healing</p> <p>DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:</p> <p>Circulation Status (NOC)</p> <p>Demonstrate improved tissue perfusion as evidenced by stabilized vital signs, strong/palpable peripheral pulses, adequate urine output, absence of pain; usual mentation; normal capillary refill; skin warm/dry; nailbeds and lips of natural pale, pink color; absence of paresthesias.</p>

ACTIONS/INTERVENTIONS	RATIONALE
<p>Circulatory Care: Arterial [or] Venous Insufficiency (NIC)</p> <p>Independent</p> <p>Monitor vital signs carefully. Assess pulses for rate, rhythm, and volume. Note hypotension; rapid, weak, thready pulse; and increased/shallow respirations.</p> <p>Assess skin for coolness, pallor, cyanosis, diaphoresis, delayed capillary refill.</p> <p>Note changes in level of consciousness; reports of headaches, dizziness; development of sensory/motor deficits (e.g., hemiparesis or paralysis), seizure activity.</p> <p>Maintain adequate fluid intake. (Refer to ND: Fluid Volume, risk for deficient, following.) Monitor urine output.</p> <p>Assess lower extremities for skin texture, edema, ulcerations (especially of internal and external malleoli).</p> <p>Investigate reports of change in character of pain, or development of bone pain, angina, tingling of extremities, eye pain/vision disturbances.</p> <p>Maintain environmental temperature and body warmth without overheating.</p> <p>Evaluate for developing edema (including genitals in men).</p>	<p>Sludging and sickling in peripheral vessels may lead to complete or partial obliteration of a vessel with diminished perfusion to surrounding tissues. Sudden massive splenic sequestration of cells can lead to shock.</p> <p>Changes reflect diminished circulation/hypoxia potentiating capillary occlusion. (Refer to ND: Gas Exchange, impaired)</p> <p>Changes may reflect diminished perfusion to the central nervous system (CNS) due to ischemia or infarction. Stagnant cells must be mobilized immediately to reduce further ischemia/infarction.</p> <p>Dehydration not only causes hypovolemia but increases sickling and occlusion of capillaries. Decreased renal perfusion/failure may occur because of vascular occlusion.</p> <p>Reduced peripheral circulation often leads to dermal changes and delayed healing.</p> <p>Changes may reflect increased sickling of cells/diminished circulation with further involvement of organs, e.g., myocardial infarction (MI) or pulmonary infarction, occlusion of vasculature of the eye.</p> <p>Prevents vasoconstriction; aids in maintaining circulation and perfusion. Excessive body heat may cause diaphoresis, adding to insensible fluid losses and risk of dehydration.</p> <p>Vaso-occlusion/circulatory stasis may lead to edema of extremities (and priapism in men), potentiating risk of tissue ischemia/necrosis.</p>
<p>Collaborative</p> <p>Monitor laboratory studies, e.g.: ABGs, CBC, LDH, AST/ALT, CPK, BUN;</p> <p>Serum electrolytes. Provide replacements as indicated.</p>	<p>Decreased tissue perfusion may lead to gradual infarction of organ tissues, such as the brain, liver, spleen, kidney, skeletal muscle, and so forth, with consequent release of intracellular enzymes.</p> <p>Electrolyte losses (especially sodium) are increased during crisis because of fever, diarrhea, vomiting, diaphoresis.</p>

ACTIONS/INTERVENTIONS	RATIONALE
<p>Circulatory Care: Arterial [or] Venous Insufficiency (NIC)</p> <p>Independent</p> <p>Administer hypo-osmolar solutions (e.g., 0.45 normal saline) via an infusion pump.</p> <p>Administer hydroxyurea (Droxia); or experimental antisickling agents (e.g., sodium cyanate) carefully. Observe for possible lethal side effects.</p> <p>Assist with/prepare for surgical diathermy or photocoagulation.</p> <p>Assist with/prepare for needle aspiration of blood from corpora cavernosa;</p> <p>Surgical intervention.</p>	<p>Hydration lowers the HbS concentration within the RBCs, which decreases the sickling tendency and also reduces blood viscosity, which helps to maintain perfusion. Infusion pump may prevent circulatory overload. <i>Note:</i> Lactated Ringer's solution or D₅W may cause RBC hemolysis and potentiate thrombus formation.</p> <p>Hydroxyurea, a cytotoxic agent, dramatically decreases the number of sickle cell episodes (by as much as 50%) and reduces the severity of complications such as fever and severe chest pain by increasing the level of fetal hemoglobin (hemoglobin F). Levels greater than 20% may prolong life. Antisickling agents (currently under investigational use) are aimed at prolonging erythrocyte survival and preventing sickling by affecting cell membrane changes. <i>Note:</i> Use of anticoagulants, plasma expanders, nitrates, vasodilators, and alkylating agents has proved essentially unsuccessful in the management of the vaso-occlusive crisis.</p> <p>Direct coagulation of bleeding sites in the eye (resulting from vascular stasis/edema) may prevent progression of proliferative changes if initiated early.</p> <p>Sickling within the penis can cause sustained erection (priapism) and edema. Removal of sludged sickled cells can improve circulation, decreasing psychological trauma and risk of necrosis/infection.</p> <p>Direct incision and ligation of the dorsal arteries of the penis and saphenocavernous shunting may be necessary in severe cases of priapism to prevent tissue necrosis.</p>

<p>NURSING DIAGNOSIS: Fluid Volume, risk for deficient</p> <p>Risk factors may include</p> <p>Increased fluid needs, e.g., hypermetabolic state/fever, inflammatory processes</p> <p>Renal parenchymal damage/infarctions limiting the kidney's ability to concentrate urine (hyposthenuria)</p> <p>Possibly evidenced by</p> <p>[Not applicable; presence of signs and symptoms establishes an <i>actual</i> diagnosis.]</p> <p>DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:</p> <p>Hydration (NOC)</p> <p>Maintain adequate fluid balance as evidenced by individually appropriate urine output with a near-normal specific gravity, stable vital signs, moist mucous membranes, good skin turgor, and prompt capillary refill.</p>
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ACTIONS/INTERVENTIONS	RATIONALE
<p>Fluid Monitoring (NIC)</p> <p>Independent</p> <p>Maintain accurate I&O. Weigh daily.</p> <p>Note urine characteristics and specific gravity.</p> <p>Monitor vital signs, comparing with patient's normal/previous readings. Take BP in lying, sitting, and standing positions if possible.</p> <p>Observe for fever, changes in level of consciousness, poor skin turgor, dryness of skin and mucous membranes, pain.</p> <p>Monitor vital signs closely during blood transfusions and note presence of dyspnea, crackles, rhonchi, wheezes, JVD, diminished breath sounds, cough, frothy sputum, and cyanosis.</p>	<p>Patient may reduce fluid intake during periods of crisis because of malaise, anorexia, and so on. Dehydration from vomiting, diarrhea, fever may reduce urine output and precipitate a vaso-occlusive crisis.</p> <p>Kidney can lose its ability to concentrate urine, resulting in excessive losses of dilute urine and fixation of the specific gravity.</p> <p>Reduction of circulating blood volume can occur from increased fluid loss, resulting in hypotension and tachycardia.</p> <p>Symptoms reflective of dehydration/hemoconcentration with consequent vaso-occlusive state.</p> <p>Patient's heart may already be weakened and prone to failure because of chronic demands placed on it by the anemic state. Heart may be unable to tolerate the added fluid volume from transfusions or rapid IV fluid administered to treat crisis/shock.</p>
<p>Collaborative</p> <p>Administer IV fluids as indicated.</p> <p>Monitor laboratory studies, e.g., Hb/Hct, serum and urine electrolytes.</p>	<p>Replaces losses/deficits; may reverse renal concentration of RBCs/presence of failure. Fluids must be given immediately (especially in CNS involvement) to decrease hemoconcentration and prevent further infarction.</p> <p>Elevations may indicate hemoconcentration. Kidneys' loss of ability to concentrate urine may result in serum depletions of Na⁺, K⁺, and Cl⁻, necessitating replacement.</p>

NURSING DIAGNOSIS: Pain, acute/chronic

May be related to

Intravascular sickling with localized stasis, occlusion, and infarction/necrosis
Activation of pain fibers due to deprivation of oxygen and nutrients, accumulation of noxious metabolites

Possibly evidenced by

Localized, migratory, or more generalized pain, described as throbbing, gnawing, or severe and incapacitating;
affecting peripheral extremities, bones, joints, back, abdomen, or head (headaches recurrent/transient)
Decreased ROM, guarding of the affected areas
Facial grimacing, narrowed/self-focus

DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:

Pain Level (NOC)

Verbalize relief/control of pain.
Demonstrate relaxed body posture, have freedom of movement, be able to sleep/rest appropriately.

ACTIONS/INTERVENTIONS	RATIONALE
<p>Pain Management (NIC)</p> <p>Independent</p> <p>Assess reports of pain, including location, duration, and intensity (scale of 0–10).</p> <p>Observe nonverbal pain cues, e.g., gait disturbances, body positioning, reluctance to move, facial expressions, and physiological manifestations of acute pain (e.g., elevated BP, tachycardia, increased respiratory rate). Explore discrepancies between verbal and nonverbal cues.</p> <p>Discuss with the patient/SO what pain relief measures were effective in the past.</p> <p>Explore alternative pain relief measures, e.g., relaxation techniques, biofeedback, yoga, meditation, progressive relaxation techniques, distraction (e.g., visual, auditory, tactile, kinesthetic, guided imagery, and breathing techniques).</p> <p>Provide support for and carefully position affected extremities.</p> <p>Apply local massage gently to affected areas.</p> <p>Encourage ROM exercises.</p> <p>Plan activities during peak analgesic effect.</p> <p>Maintain adequate fluid intake.</p>	<p>Sickling of cells potentiates cellular hypoxia and may lead to infarction of tissues with resultant pain. Typically pain occurs in the back, ribs, and limbs and lasts 5–7 days.</p> <p>Pain is unique to each patient; therefore, one may encounter varying descriptions because of individualized perceptions. Nonverbal cues may aid in evaluation of pain and effectiveness of therapy.</p> <p>Involves patient/SO in care and allows for identification of remedies that have already been found to relieve pain. Helpful in establishing individualized treatment needs.</p> <p>Cognitive-behavioral interventions may reduce reliance on pharmacological therapy and enhance patient’s sense of control.</p> <p>Reduces edema, discomfort, and risk of injury, especially if osteomyelitis is present.</p> <p>Helps reduce muscle tension.</p> <p>Prevents joint stiffness and possible contracture formation.</p> <p>Maximizes movement of joints, enhancing mobility.</p> <p>Dehydration increases sickling/vaso-occlusion and corresponding pain.</p>

ACTIONS/INTERVENTIONS	RATIONALE
<p>Pain Management (NIC)</p> <p>Collaborative</p> <p>Apply warm, moist compresses to affected joints or other painful areas. Avoid use of ice or cold compresses.</p> <p>Administer medications as indicated: narcotics, e.g., continuous infusion around-the-clock morphine (Astramorph, Duramorph), hydromorphone (Dilaudid); nonnarcotic analgesics, e.g., acetaminophen (Tylenol); or sedatives, e.g., hydroxyzine (Vistaril).</p> <p>Administer/monitor RBC transfusion.</p>	<p>Warmth causes vasodilation and increases circulation to hypoxic areas. Cold causes vasoconstriction and compounds the crisis.</p> <p>Reduces pain and promotes rest and comfort. <i>Note:</i> Narcotics are the mainstay of pain control during crisis. Acetaminophen (Tylenol) can be used for control of headache, pain, and fever. Aspirin should be avoided because it alters blood pH and can make cells sickle more easily. Meperidine (Demerol) should not be used because its metabolite, normeperidine, can cause CNS excitation, e.g., anxiety, tremors, seizures.</p> <p>Frequency of painful crises may be reduced by routine partial exchange transfusions to maintain population of normal RBCs.</p>

<p>NURSING DIAGNOSIS: Mobility, impaired physical</p> <p>May be related to</p> <p>Multiple/recurrent bone infarctions or infections (weight-bearing bones) Pain/discomfort: kyphosis of upper back/lordosis of lower back, possible joint effusions Osteoporosis with fragmentation/collapse of femoral head or vertebra (compression deformities) Bacterial infections (osteomyelitis)</p> <p>Possibly evidenced by</p> <p>Reports of pain Limited joint ROM, reluctance to move, inability to walk/perform ADLs, guarding of joints, gait disturbances Generalized weakness, therapeutic restrictions (e.g., bedrest)</p> <p>DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:</p> <p>Mobility Level (NOC)</p> <p>Maintain/increase strength and function of affected body parts. Participate in activities with absence of or improvement in gait disturbances, increased joint ROM, and absence of inflammatory signs.</p>
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Refer to CPs: Extended Care, ND: Mobility, impaired physical.

NURSING DIAGNOSIS: Skin integrity, risk for impaired

Risk factors may include

Impaired circulation (venous stasis and vaso-occlusion); altered sensation
Decreased mobility/bedrest

Possibly evidenced by

[Not applicable; presence of signs and symptoms establishes an *actual* diagnosis.]

DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:

Tissue Integrity: Skin and Mucous Membranes (NOC)

Prevent dermal ischemic injury.
Display improvement in wound/lesion healing if present.

Risk Control (NOC)

Participate in behaviors to reduce risk factors/skin breakdown.

ACTIONS/INTERVENTIONS	RATIONALE
Skin Surveillance (NIC)	
Independent	
Reposition frequently, even when sitting in chair.	Prevents prolonged tissue pressure where circulation is already compromised, reducing risk of tissue trauma/ischemia.
Inspect skin/pressure points regularly for redness, provide gentle massage.	Poor circulation may predispose to rapid skin breakdown.
Protect bony prominences with sheepskin, heel/elbow protectors, pillows, as indicated.	Decreases pressure on tissues, preventing skin breakdown.
Keep skin surfaces dry and clean; linens dry/wrinkle-free. Monitor ischemic areas, leg bruises, cuts, bumps closely for ulcer formation.	Moist, contaminated areas provide excellent media for growth of pathogenic organisms. Potential entry sites for pathogenic organisms. In presence of altered immune system, this increases risk of infection/delayed healing.
Elevate lower extremities when sitting.	Enhances venous return, reducing venous stasis/edema formation.
Collaborative	
Provide egg-crate, alternating air pressure, or water mattress.	Reduces tissue pressure and aids in maximizing cellular perfusion to prevent dermal injury.
Cleanse open wounds/ulcers with hydrogen peroxide, boric acid, or povidone iodine (Betadine) solutions as indicated. Monitor distribution, size, depth, character, and drainage.	Improvement or delayed healing reflects status of tissue perfusion and effectiveness of interventions. <i>Note:</i> These patients are at increased risk of serious complications because of lowered resistance to infection and decreased nutrients for healing.
Prepare for/assist with hyperbaric oxygenation to ulcer sites.	Maximizes oxygen delivery to tissues, enhancing healing.

NURSING DIAGNOSIS: Infection, risk for

Risk factors may include

Chronic disease process, tissue destruction, e.g., infarction, fibrosis, loss of spleen (autosplenectomy)
Inadequate primary defenses (broken skin, stasis of body fluids, decreased ciliary action)

Possibly evidenced by

[Not applicable; presence of signs and symptoms establishes an *actual* diagnosis]

DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:

Knowledge: Infection Control (NOC)

Verbalize understanding of individual causative/risk factors.
Identify interventions to prevent/reduce risk of infection.

Refer to CPs: Pneumonia: Microbial, Sepsis/Septicemia; Fractures, ND: Infection, risk for.

NURSING DIAGNOSIS: Knowledge, deficient [Learning Need] regarding condition, prognosis, treatment, self-care, and discharge needs

May be related to

Lack of exposure/recall
Information misinterpretation
Unfamiliarity with resources

Possibly evidenced by

Questions, request for information, statement of misconceptions
Inaccurate follow-through of instructions; development of preventable complications
Verbal/nonverbal cues of anxiety

DESIRED OUTCOMES/EVALUATION CRITERIA—PATIENT WILL:

Knowledge: Disease Process (NOC)

Verbalize understanding of disease process, including symptoms of crisis, potential complications.

Knowledge: Treatment Regimen (NOC)

Verbalize understanding of therapeutic needs.
Initiate necessary behaviors/lifestyle changes to prevent complications.
Participate in continued medical follow-up; genetic counseling/family planning services.

ACTIONS/INTERVENTIONS

Teaching: Disease Process (NIC)

Independent

Review disease process and treatment needs.

RATIONALE

Provides knowledge base from which patient can make informed choices. *Note:* The median age at death for females is 48 yr, and 42 yr for males, with death often due to organ failure.

ACTIONS/INTERVENTIONS	RATIONALE
<p>Teaching: Disease Process (NIC)</p>	
<p>Independent</p>	
<p>Review precipitating factors, e.g.: Cold environmental temperatures, failure to dress warmly when engaging in winter activities; wearing tight, restrictive clothing; stressful situations;</p>	<p>Causes peripheral vasoconstriction, which may result in sludging of the circulation, increased sickling, and may precipitate a vaso-occlusive crisis.</p>
<p>Strenuous physical activity/contact-type sports, and extremely warm temperatures;</p>	<p>Increases metabolic demand for oxygen and increases insensible fluid losses (evaporation and perspiration) leading to dehydration, which may increase blood viscosity and tendency to sickle.</p>
<p>Travel to places more than 7000 ft above sea level or flying in unpressurized aircraft;</p>	<p>Decreased oxygen tension present at higher altitudes causes hypoxia and potentiates sickling of cells.</p>
<p>Encourage consumption of at least 4–6 qt of fluid daily, during a steady state of the disease, increasing to 6–8 qt during a painful crisis or while engaging in activities that might precipitate dehydration.</p>	<p>Prevents dehydration and consequent hyperviscosity that can potentiate sickling/crisis.</p>
<p>Encourage ROM exercise and regular physical activity with a balance between rest and activity.</p>	<p>Prevents bone demineralization and may reduce risk of fractures. Aids in maintaining level of resistance and decreases oxygen needs.</p>
<p>Review patient’s current diet, reinforcing the importance of diet including liver, green leafy vegetables, citrus fruits, and wheat germ. Provide necessary instruction regarding supplementary vitamins such as folic acid.</p>	<p>Sound nutrition is essential because of increased demands placed on bone marrow (e.g., folate and vitamin B₁₂ are used in greater quantities than usual). Folic acid supplements are frequently ordered to prevent aplastic crisis.</p>
<p>Discourage smoking and alcohol consumption; identify appropriate community support groups.</p>	<p>Nicotine induces peripheral vasoconstriction and decreases oxygen tension, which may contribute to cellular hypoxia and sickling. Alcohol increases the possibility of dehydration (precipitating sickling). Maintaining these changes in behavior/lifestyle may require prolonged support.</p>
<p>Discuss principles of skin/extremity care and protection from injury. Encourage prompt treatment of cuts, insect bites, sores.</p>	<p>Because of impaired tissue perfusion, especially in the periphery, distal extremities are especially susceptible to altered skin integrity/infection.</p>
<p>Include instructions on care of leg ulcers that might develop.</p>	<p>Fosters independence and maintenance of self-care at home.</p>
<p>Instruct patient to avoid persons with infections such as upper respiratory infections (URIs).</p>	<p>Altered immune response places patient at risk for infections, especially bacterial pneumonia.</p>
<p>Recommend patient avoid cold remedies and decongestants containing ephedrine and large amounts of caffeine. Stress the importance of reading labels on over-the-counter (OTC) drugs and consulting healthcare provider before consuming any drugs/herbal supplements.</p>	<p>Those remedies containing vasoconstrictors may decrease peripheral tissue perfusion and cause sludging of sickled cells.</p>

ACTIONS/INTERVENTIONS	RATIONALE
<p>Teaching: Disease Process (NIC)</p>	
<p>Independent</p>	
<p>Discuss conditions for which medical attention should be sought, e.g.:</p>	
<p>Urine that appears blood tinged or smoky;</p>	<p>Symptoms suggestive of sickling in the renal medulla.</p>
<p>Indigestion, persistent vomiting, diarrhea, high fever, excessive thirst;</p>	<p>Dehydration may trigger a vaso-occlusive crisis.</p>
<p>Severe joint or bone pain;</p>	<p>May signify a vaso-occlusive crisis due to sickling in the bones or spleen (ischemia or infarction) or onset of osteomyelitis.</p>
<p>Severe chest pain, with or without cough;</p>	<p>May reflect angina, impending MI, or pneumonia.</p>
<p>Abdominal pain; gastric distress following meals;</p>	<p>Cholelithiasis, primarily with bilirubin stones, is present in more than 50% of adults.</p>
<p>Fever, swelling, redness, increasing fatigue/pallor, dizziness, drowsiness, nonhealing leg ulcers.</p>	<p>Suggestive of infections that may precipitate a vaso-occlusive crisis if dehydration develops. <i>Note:</i> Severe infections are the most frequent cause of aplastic crisis.</p>
<p>Assist patient to strengthen coping abilities, e.g., deal appropriately with anxiety, get adequate information, use relaxation techniques.</p>	<p>Promotes patient's sense of control, may avert a crisis.</p>
<p>Suggest wearing a medical alert bracelet or carrying a wallet card.</p>	<p>May prevent inappropriate treatment in emergency situation.</p>
<p>Discuss genetic implications of the condition. Encourage SO/family members to seek testing to determine presence of HbS.</p>	<p>Screening may identify other family members with sickle cell trait. Hereditary nature of the disease with the possibility of transmitting the mutation may have a bearing on the decision to have children.</p>
<p>Explore concerns regarding childbearing/family planning and refer to community resources and obstetrician knowledgeable about sickle cell disease, as indicated.</p>	<p>Provides opportunity to correct misconceptions/present information necessary to make informed decisions. Pregnancy can precipitate a vaso-occlusive crisis because the placenta's tortuous blood supply and low oxygen tension potentiate sickling, which in turn can lead to fetal hypoxia.</p>
<p>Encourage patient to have routine follow-ups, e.g.:</p> <p>Periodic laboratory studies, e.g., CBC;</p>	<p>Monitors changes in blood components; identifies need for changes in treatment regimen. When using hydroxyurea, frequent monitoring of CBC is required because of narrow margin between efficacy (acceptable degree of bone marrow suppression) and toxicity (neutropenia, anemia, thrombocytopenia).</p>
<p>Biannual dental examination;</p>	<p>Sound oral hygiene limits opportunity for bacterial invasion/sepsis.</p>
<p>Annual ophthalmologic examination.</p>	<p>Detects development of sickle retinopathy with either proliferative or nonproliferative ocular changes.</p>

ACTIONS/INTERVENTIONS	RATIONALE
<p data-bbox="235 279 610 306">Teaching: Disease Process (NIC)</p> <p data-bbox="235 325 397 352">Independent</p> <p data-bbox="235 371 695 399">Determine need for vocational/career guidance.</p> <p data-bbox="235 489 766 600">Encourage participation in community support groups available to sickle cell patients/SO, such as the Sickle Cell Disease Association of America, March of Dimes, public health/visiting nurse.</p>	<p data-bbox="820 371 1325 453">Sedentary career may be necessary because of the decreased oxygen-carrying capacity and diminished exercise tolerance.</p> <p data-bbox="820 489 1377 632">Helpful in adjustment to long-term situation; reduces feelings of isolation and enhances problem solving through sharing of common experiences. <i>Note:</i> Failure to resolve concerns/deal with situation may require more intensive therapy/psychological support.</p>

POTENTIAL CONSIDERATIONS following acute hospitalization (dependent on patient's age, physical condition/presence of complications, personal resources, and life responsibilities)

Pain, acute/chronic—intravascular sickling with localized stasis, occlusion, and infarction/necrosis; activation of pain fibers due to deprivation of oxygen and nutrients, accumulation of noxious metabolites.

Fluid Volume, risk for deficient—increased fluid needs, e.g., hypermetabolic state/fever, inflammatory processes; renal parenchymal damage/infarctions limiting the kidney's ability to concentrate urine (hyposthenuria).

Infection, risk for—chronic disease process, tissue destruction, e.g., infarction, fibrosis, loss of spleen (autosplenectomy); inadequate primary defenses (broken skin, stasis of body fluids, decreased ciliary action).