DIANE'S NGN NCLEX 2026 STUDY SERIES

Sickle Cell Disease

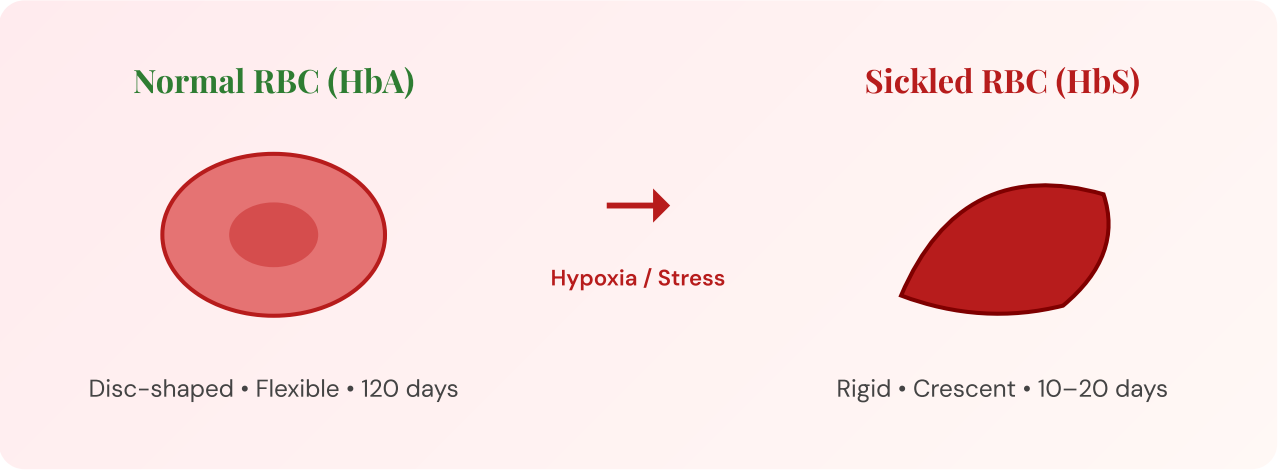
An inherited hemoglobinopathy producing rigid, crescent-shaped RBCs that cause vaso-occlusion, chronic hemolytic anemia, and life-threatening organ ischemia.

Hematology · Genetics · Oncology Adjacent

Source note: Sickle cell content was not in your uploaded study notes — built from standard NGN NCLEX 2026 nursing references (Saunders, ATI, Hogan, Lewis Med-Surg).

1 Definition / Overview

- **Autosomal recessive** genetic disorder — both parents must carry the trait. Child needs **two copies** (HbSS) to have the disease; **one copy** = sickle cell trait (carrier, usually asymptomatic).
- Abnormal **hemoglobin S (HbS)** replaces normal HbA. Under stress (low O_2, dehydration, acidosis), HbS polymerizes and RBCs deform into rigid sickle/crescent shapes.
- Sickled cells **occlude small vessels** → tissue ischemia, severe pain, organ damage. Lifespan of sickled RBCs drops from 120 days to **10–20 days** → chronic hemolytic anemia.
- Highest prevalence: African, Mediterranean, Middle Eastern, Caribbean, and Indian descent. Diagnosed by **newborn screening** + **hemoglobin electrophoresis** (definitive).



2 Pathophysiology (Simplified)

1. **Trigger** occurs (hypoxia, dehydration, infection, cold, acidosis, stress, high altitude)

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2. **HbS polymerizes** when oxygen drops — hemoglobin chains stack into rigid rods inside the RBC

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3. RBC **deforms into sickle shape** — rigid, sticky, fragile

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4. Sickled cells **clump and obstruct microvasculature** → vaso-occlusion → tissue ischemia → severe pain + organ damage

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5. Cells **hemolyze prematurely** (10–20 day lifespan) → chronic anemia, jaundice, gallstones, fatigue

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6. Repeated crises → **cumulative organ damage**: spleen autoinfarcts, kidneys fail, retinas detach, strokes, AVN of hips

🧯 **The 6 H/A/I/D Triggers (HAIDCS):** **H** ypoxia · **A** cidosis · **I** nfection · **D** ehydration · **C** old · **S** tress (physical/emotional)

3 Signs & Symptoms — by Crisis Type

Crisis Type	Hallmark Findings	Red Flag 🚨
Vaso-Occlusive (VOC) Most common	Severe deep bone/joint pain, abdominal pain, swelling of hands & feet (**dactylitis** in infants), fever, priapism	Pain rated 8–10/10 — treat aggressively
Acute Chest Syndrome #1 cause of death	Chest pain, fever, cough, dyspnea, hypoxia, new pulmonary infiltrates on CXR	🚨 LIFE-THREATENING — mimics PE/pneumonia
Splenic Sequestration Children	Sudden splenomegaly, LUQ pain, hypotension, shock, profound drop in Hgb	🚨 Hypovolemic shock — emergency transfusion
Aplastic Crisis	Severe anemia, pallor, fatigue, low reticulocyte count — often triggered by parvovirus B19	Bone marrow failure — needs transfusion
Hemolytic Crisis	Jaundice, scleral icterus, dark urine, drop in Hgb, elevated bilirubin	Risk of gallstones, hyperbilirubinemia

⏰ Late / Chronic Complications

Neuro
Stroke (esp. children 2–16 yrs), TIA, cognitive deficits

Renal
Hyposthenuria (can't concentrate urine), CKD, hematuria

Ocular
Retinopathy, retinal detachment, vision loss

MSK
Avascular necrosis (AVN) of hip, osteomyelitis (Salmonella)

Skin
Chronic leg ulcers (especially malleoli)

Cardiac
Cardiomegaly, pulmonary HTN, heart failure

4 Priority Nursing Interventions (ABC + HOP)

Airway / Breathing / Circulation

- **Assess respiratory status** — RR, SpO_2, breath sounds. New cough/dyspnea/chest pain = **acute chest syndrome** until proven otherwise.
- **Administer humidified oxygen** to keep SpO_2 ≥ 95% (oxygen reverses sickling).
- **Monitor for shock** in children — sudden splenomegaly + hypotension = sequestration.

The HOP Priorities

H — Hydration

IV fluids @ 1.5× maintenance + PO fluids. Reverses RBC dehydration and dilutes sickled cells.

O — Oxygenation

Humidified O_2 to maintain SpO_2 ≥ 95%. Treats hypoxia driving sickling.

P — Pain Control

IV opioids (morphine, hydromorphone) on a schedule + PCA. **NEVER meperidine.**

Additional Priorities

- **Warmth** — apply warm compresses to painful areas. **NEVER ice/cold** (worsens vasoconstriction → worse sickling).
- **Rest** — bed rest during acute crisis to reduce O_2 demand.
- **Infection prevention** — strict hand hygiene, prompt antibiotics for fever (sickle cell pts are functionally asplenic = high sepsis risk).
- **Blood transfusions** — for severe anemia, acute chest syndrome, stroke, or pre-op. Watch for iron overload long-term.
- **Monitor labs** — Hgb (target 7–9 g/dL; higher can worsen viscosity), reticulocyte count, bilirubin, LDH.
- **Daily weights, strict I/Os** — track fluid balance closely.

5 Pharmacology

Drug / Class	Mechanism	Key Side Effects / Nursing Notes
Hydroxyurea (Droxia, Hydrea)	↑ Fetal hemoglobin (HbF) production → less HbS polymerization → fewer crises	Bone marrow suppression — monitor CBC. **Teratogenic** — contraception required. Takes weeks for effect.
Voxelotor (Oxbryta)	Binds HbS → keeps it oxygenated → prevents polymerization	Headache, diarrhea, nausea. Approved age ≥ 4 yrs.
Crizanlizumab (Adakveo)	P-selectin inhibitor — blocks RBC adhesion to vessel walls → fewer VOCs	IV monthly. Watch for infusion reactions.
L-glutamine (Endari)	Reduces oxidative stress on RBCs	Oral powder; mix with food/drink. ↓ frequency of crises.
Penicillin V prophylaxis	Prevents pneumococcal sepsis (high risk d/t functional asplenia)	**Children < 5 yrs daily**, sometimes longer. Critical for prevention.
Folic acid	Supports rapid RBC turnover	Daily PO. Inexpensive but essential.
Morphine / Hydromorphone	Opioid analgesia for VOC pain	IV scheduled or PCA. Monitor RR, sedation. Constipation prophylaxis.
✕ **Meperidine (Demerol)**	Opioid — **CONTRAINDICATED**	Metabolite normeperidine accumulates → **seizures**. Never give for sickle cell pain.

💉 Required Vaccinations

Pneumococcal (PCV13 + PPSV23) · Meningococcal · Hib · Annual influenza · Hepatitis B · COVID-19.
Sickle cell patients are **functionally asplenic** — encapsulated organisms are deadly.

6 NCLEX Test Traps ⚠️

⚠ ~~"Apply ice packs to the painful joint."~~
✓ Apply **warm** compresses. Cold causes vasoconstriction → worsens sickling.

⚠ ~~"Administer meperidine (Demerol) for severe pain."~~
✓ Use **morphine or hydromorphone**. Meperidine's metabolite causes seizures in sickle cell.

⚠ ~~"Restrict fluids to reduce edema."~~
✓ **Increase** fluids (1.5× maintenance). Dehydration drives sickling.

⚠ ~~"This patient is drug-seeking — wait before giving pain meds."~~
✓ Sickle cell pain is **real, severe, and undertreated**. Treat aggressively per protocol — bias kills.

⚠ ~~"Both parents must have the disease for a child to inherit it."~~
✓ Both parents must carry the **trait** (one HbS gene). Two carriers = 25% chance of disease per pregnancy.

⚠ ~~"Tourniquets and BP cuffs are fine on a sickle cell patient."~~
✓ Avoid **prolonged tourniquet use** — local hypoxia triggers sickling. Limit BP cuff inflation time.

⚠ ~~"A new cough and chest pain in a sickle cell patient is just anxiety."~~
✓ **Acute Chest Syndrome** — leading cause of death. Notify provider immediately, get CXR & ABG.

7 Patient & Family Education

🚫 Avoid Triggers

- **High altitudes** & unpressurized aircraft (low O_2)
- Extreme cold — dress warmly, avoid swimming in cold water
- **Smoking, alcohol, dehydration**
- Strenuous exercise to exhaustion
- Stressful situations when possible

✅ Daily Self-Management

- Drink **2–3 L water/day** (more in heat or illness)
- Take prescribed meds: hydroxyurea, folic acid, penicillin (kids)
- Stay current on **all vaccinations**
- Prompt fever treatment — fever > 101°F = ER visit
- Wear a **medical alert bracelet**

🧬 **Genetic counseling** — recommend for any couple where both partners are carriers (25% chance of disease, 50% trait, 25% unaffected per pregnancy). Newborn screening identifies it at birth.

🚨 **When to call 911 / go to ER:** Fever ≥ 101°F · Chest pain or trouble breathing · Severe headache or neurologic changes (stroke) · Sudden left-sided abdominal pain (sequestration) · Priapism > 4 hours · Vision changes · Pain not relieved by home meds.

8 Memory Boosters & Mnemonics

💧 "HOP" — The 3 Sickle Cell Crisis Priorities

H ydration (IV + PO fluids)

O xygenation (humidified O_2)

P ain control (morphine/hydromorphone — **NOT** meperidine)

🚨 "HAIDCS" — The Sickle Crisis Triggers

H ypoxia · **A** cidosis · **I** nfection · **D** ehydration · **C** old · **S** tress

❌ "Demerol Drops the Threshold"

Meperidine (Demerol) is contraindicated in sickle cell — its metabolite `normeperidine` drops the seizure threshold. Pick **morphine** instead.

🥶 "Warm = Welcome, Cold = Crisis"

Always apply **warm** compresses to painful areas. **NEVER cold** — vasoconstriction worsens sickling.

🫁 "ACS = Acute Chest = Always Critical"

Any sickle cell patient with new fever + chest pain + cough + dyspnea = **Acute Chest Syndrome**. #1 cause of death — notify provider STAT.

9 Key Labs & Diagnostics

Test	Expected Finding	Why It Matters
Hgb electrophoresis	HbSS > 80% (disease) or HbAS (trait)	**Definitive diagnosis** — identifies HbS
Newborn screening	Mandatory in all 50 states	Early detection → early prophylactic penicillin
Sickledex (sickle solubility test)	Positive	Quick screen but does NOT distinguish trait from disease
CBC	Hgb 6–9 g/dL, ↑ retic count, ↑ WBC	Chronic hemolytic anemia baseline
Peripheral smear	Sickle cells, target cells, Howell-Jolly bodies	Howell-Jolly = functional asplenia
Bilirubin (indirect)	Elevated	From hemolysis — explains jaundice/gallstones
Transcranial Doppler (TCD)	↑ velocity = stroke risk	Annual screening in children 2–16 yrs
Chest X-ray	New infiltrate	Diagnoses Acute Chest Syndrome

10 Life-Threatening Complications

🫁 Acute Chest Syndrome

Fever, chest pain, cough, dyspnea, hypoxia, new CXR infiltrate. **#1 cause of death.** Treat with O_2, antibiotics, transfusion, hydroxyurea.

🧠 Stroke

Most common in children 2–16 yrs. Sudden weakness, slurred speech, vision changes, severe headache. Annual TCD screening prevents.

🩸 Splenic Sequestration

Pediatric emergency. Sudden splenomegaly + drop in Hgb + shock. Needs immediate transfusion.

⚡ Sepsis

Functional asplenia → encapsulated organism risk (S. pneumo, H. flu, N. meningitidis). Any fever in a sickle cell child = ER.

🍆 Priapism

Sustained painful erection > 4 hours. Urologic emergency — can cause permanent impotence.

🦴 AVN of the Hip

Avascular necrosis from chronic vaso-occlusion. May require hip replacement.

11 NCJMM Clinical Judgment Scenario

Scenario: *A 9-year-old with sickle cell disease (HbSS) is admitted to the pediatric unit with severe leg and back pain rated 9/10. Vitals: HR 132, RR 28, BP 102/64, SpO_2 92% on room air, Temp 100.8°F (38.2°C). The child is crying, splinting, and refusing fluids. Mom reports the child played outside in cold weather yesterday and has had a runny nose for 3 days. Hgb is 7.2 g/dL (baseline 8.5).*

1 Recognize Cues

Severe pain (9/10), tachycardia, tachypnea, **SpO_2 92%**, low-grade fever, dehydration risk (refusing fluids), recent cold exposure + URI symptoms, drop in Hgb from baseline.

2 Analyze Cues

Multiple sickling triggers stacked: **cold + infection + dehydration + hypoxia**. Pain pattern + drop in Hgb consistent with vaso-occlusive crisis. Low SpO_2 + tachypnea + fever raise concern for evolving **acute chest syndrome**.

3 Prioritize Hypotheses

① Vaso-occlusive crisis (highest probability) → ② Possible early acute chest syndrome (highest acuity) → ③ Underlying viral infection driving crisis. **ACS must be ruled out first** — leading cause of death.

4 Generate Solutions

Apply humidified O_2 to maintain $SpO_2 \geq 95\%$ · Establish IV access for fluids at 1.5× maintenance · Administer scheduled IV morphine (NOT meperidine) · Obtain CXR, blood cultures, CBC with retic, type & screen · Apply warm compresses · Notify provider for ACS workup.

5 Take Action

Priority order: ① Apply O_2 (airway/breathing) → ② Start IV NS at 1.5× maintenance → ③ Administer ordered IV morphine → ④ Obtain CXR & labs → ⑤ Warm compress to painful areas → ⑥ Reassess pain & SpO_2 in 15–30 min → ⑦ Notify provider of any worsening.

6

Evaluate Outcomes

Effective: Pain ≤ 3/10, SpO_2 ≥ 95%, HR/RR returning to age-normal, child taking PO fluids, afebrile, CXR clear. **Ineffective / Escalate:** Worsening dyspnea, new infiltrate on CXR, SpO_2 < 92% on O_2, neurologic changes, Hgb continuing to drop → transfer to PICU, prepare for exchange transfusion.