Sickle Cell *Disease*

A clinical-judgment study poster for the 2026 Next Generation NCLEX — recognize cues, prioritize hypotheses, take action, evaluate outcomes.

01 / PATHO

What's happening *inside the cell*

An inherited autosomal-recessive hemoglobinopathy (HbSS) in which abnormal hemoglobin S polymerizes under stress — turning soft biconcave RBCs into rigid crescent-shaped sickles that clog microvasculature, hemolyze early, and starve tissues of oxygen.

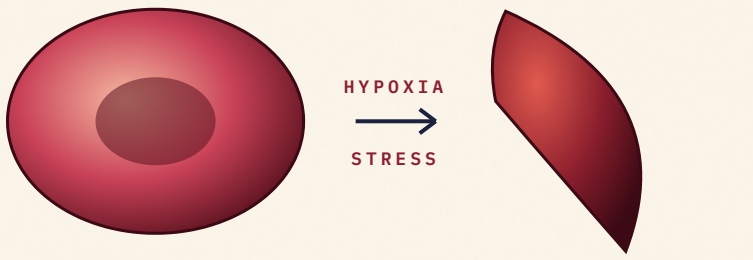
Key concept

Under **hypoxia, dehydration, acidosis, cold, stress, or infection**, HbS polymerizes. RBCs stiffen into sickles → vaso-occlusion → ischemic pain + chronic hemolytic anemia. Sickled cells live ~10–20 days (normal: 120 days).

Inheritance: Autosomal recessive. Two carrier parents = 25% disease, 50% trait, 25% unaffected. Screening is universal in U.S. newborns.

Onset: Symptoms typically emerge at **4–6 months**, when fetal hemoglobin (HbF) declines.

HBSS | HBF PROTECTIVE | CHRONIC HEMOLYSIS



NORMAL · HbA
Flexible · biconcave · 120-day life

SICKLE · HbS
Rigid · crescent · 10–20-day life

02 / CRISES

The four sickle *crises* to know

NGN items will ask you to distinguish which crisis a child is experiencing based on cues. Memorize the distinguishing finding in each.

MOST COMMON

TYPE · PAIN CRISIS

Vaso-Occlusive (VOC)

- Severe bone/joint/abdominal pain
- **Dactylitis** in infants (hand-foot syndrome) — often the *first* sign
- Fever, swelling, ↓ ROM
- Priapism in males

LIFE-THREATENING

TYPE · SEQUESTRATION

Splenic Sequestration

- Blood **pools in spleen** → hypovolemic shock
- Rapidly **enlarging spleen** + pallor
- ↓ BP, tachycardia, weak pulse
- Age 6 mo–5 yr most at risk

PARVO B19

TYPE · APLASTIC

Aplastic Crisis

- Bone marrow **stops producing** RBCs
- Triggered by **parvovirus B19** ("fifth disease")
- Profound anemia, ↓ reticulocytes
- Fatigue, pallor, tachycardia

#1 KILLER

TYPE · PULMONARY

Acute Chest Syndrome

- Chest pain, fever ≥38.5°C
- Cough, dyspnea, hypoxia (↓ SpO_2)
- **New infiltrate on CXR**
- Leading cause of death in SCD

03 / TRIGGERS

What *sets off* a sickling crisis

MNEMONIC · MEMORIZE

5 H's

Hypoxia
Low O_2 — most common trigger

Dehydration
↑ blood viscosity → sickling

Cold / Heat
Vasoconstriction & dehydration

Hypoxia · Hydration (lack of) · Heat/cold extremes · High altitude · Hyperexertion

Add **Infection** and **Acidosis** — they are the most tested precipitants on NGN stems.

High altitude
Flights, mountains = low O_2

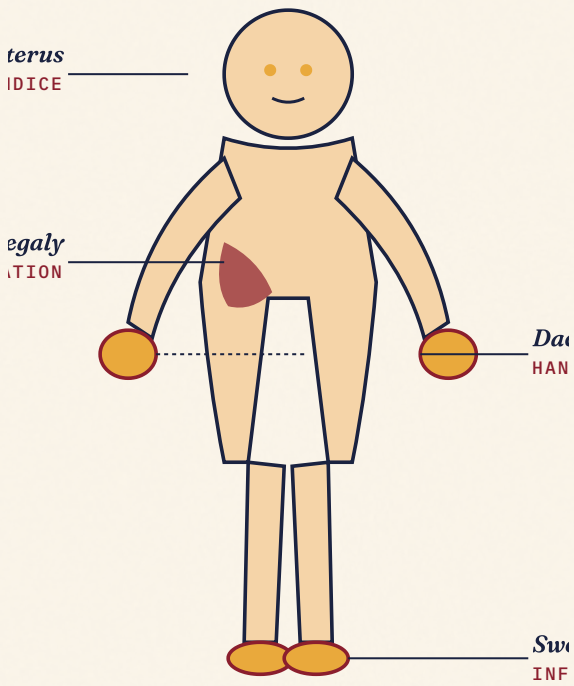
Infection
Fever = prioritize immediately

Hyperexertion
Strenuous play/sports

04 / CUES

Recognize the *clinical cues*

NGN "Recognize Cues" items will ask you to flag significant findings in a case. Know age-specific presentations — infants look different from school-age children.



1. **Dactylitis**
Painful, swollen hands/feet — often the first sign in infants 6–12 mo

2. **Pallor + fatigue**
Chronic hemolytic anemia; Hgb typically 6–9 g/dL

3. **Jaundice / icterus**
Bilirubin ↑ from rapid RBC breakdown

4. **Splenomegaly**
Then autosplenectomy by school-age → infection risk

5. **Severe pain**
Bones, joints, abdomen, chest, back — use age-appropriate pain scale

6. **Delayed growth**
Short stature, delayed puberty; chronic anemia + ↑ metabolic demand

7. **Priapism**
Prolonged painful erection — urologic emergency

8. **Stroke risk**
Screen ages 2–16 yr with **transcranial Doppler (TCD)**

05 / ACTIONS

Priority nursing care — *"HHOP"*

These four pillars underpin every take-action NGN item on a sickling child. Commit them to memory in this order.

H • H • O • P — *the sickle crisis bundle*

PRIORITIZE · IMPLEMENT · EVALUATE

H
Hydration
- IV fluids at **1.5× maintenance**
- Push PO fluids when tolerated
- Avoid fluid overload → pulm. edema
- Strict I&O, daily weights

H
Hypoxia prevention
- O_2 to keep $SpO_2 \geq$ **95%**
- Semi-Fowler's positioning
- Incentive spirometry q2h
- Monitor for acute chest syndrome

O
Opioid pain control
- **Morphine** is drug of choice
- Scheduled around-the-clock + PCA
- Ibuprofen / acetaminophen adjunct
- **Warm** compresses — never cold

P
Prevent infection
- **Fever ≥ 38.5°C = emergency**
- Blood cultures → IV antibiotics STAT
- Handwashing, sick-contact avoidance
- Keep vaccines up to date

⚠ AVOID MEPERIDINE (DEMEROL) — metabolite normeperidine lowers seizure threshold. COLD COMPRESSES — trigger vasoconstriction. TOURNIQUETS / BP CUFFS LEFT INFLATED. RESTRICTING FLUIDS.

06 / MEDS & LABS

Medications & diagnostic *data*

Hydroxyurea
DISEASE MODIFIER
Increases fetal hemoglobin (HbF) → fewer crises. Monitor CBC; teratogenic.

Penicillin V prophylaxis
INFECTION PREVENTION
Start at **2 months**

Diagnostic & monitoring labs

HGB ELECTROPHORESIS	Gold-standard diagnosis — identifies HbS, HbF, HbA

, continue through at least **age 5**. Prevents pneumococcal sepsis.

Folic acid
SUPPORTIVE

Daily — supports rapid RBC production from chronic hemolysis.

Vaccines (full schedule)
PROPHYLAXIS

PCV13 + PPSV23, meningococcal, Hib, annual **influenza**, COVID-19.

Morphine (IV/PCA)
PAIN MGMT · ACUTE

First-line opioid for crisis pain. Monitor RR, sedation. **Not meperidine.**

Transfusion / Exchange
SEVERE CRISIS

For acute chest, stroke, severe anemia. Watch for iron overload; may need chelation.

SICKLEDEX	Screening test — does not differentiate trait vs disease
NEWBORN SCREEN	Universal in all U.S. states
CBC	Hgb 6–9 g/dL · ↑ WBC · reticulocytes ↑
BILIRUBIN	Elevated (indirect) from hemolysis
TCD ULTRASOUND	Annual stroke screen, ages 2–16 yr
PULSE OXIMETRY	Trend SpO_2 — new desaturation = red flag

NGN TIP

Match the lab change to the crisis:
↓ RETICS + ↓ HGB = APLASTIC

.

↑ RETICS + RAPIDLY DROPPING HGB + BIG SPLEEN = SEQUESTRATION.

“A child with sickle cell disease and a fever ≥ 38.5 °C is always an emergency — culture, antibiotics, admit. Never wait.”

NGN CLINICAL JUDGMENT · PRIORITY CUE

07 / EDUCATION

Family & patient *teaching*

Daily living
- Drink water consistently — set reminders
- Dress warmly in cold; avoid overheating
- Avoid high altitudes & unpressurized flights
- Rest between strenuous activities
- No smoking / secondhand smoke exposure

Infection prevention
- Handwashing — entire household
- Keep ALL vaccines current
- Daily penicillin as prescribed
- Call HCP for **fever ≥ 38.5°C** immediately
- Avoid crowds during flu season

When to seek care
- Fever, chest pain, trouble breathing
- New or severe pain not relieved at home
- Sudden pallor, weakness, enlarging abdomen
- Stroke signs: facial droop, weakness, slurred speech
- Priapism > 2 hours

NGN FOCUS

Clinical judgment model — applied to sickle cell

STEP 01

Recognize & Analyze Cues

Flag: **fever, ↓ SpO_2, chest pain, enlarging spleen, severe pain, neuro changes.** Match cues to likely crisis (VOC vs acute chest vs sequestration vs aplastic).

STEP 02

Prioritize Hypotheses & Generate Solutions

Airway/breathing first: **acute chest > sequestration > VOC**. Plan the HHOP bundle: fluids, O_2, analgesia, cultures + antibiotics if febrile.

STEP 03

Take Action & Evaluate Outcomes

Implement interventions; **reassess pain, SpO_2, VS, perfusion**. Desired outcome: pain ≤ 3/10, SpO_2 ≥ 95%, HR/BP WNL, adequate UO, no new cues.

High-Yield *NCLEX* • Pediatric Hematology Poster

FOR EDUCATIONAL REVIEW ONLY · ALWAYS FOLLOW FACILITY PROTOCOL & CURRENT EVIDENCE

2026 NGN TEST PLAN
POSTER Nº 07 · SICKLE CELL
PRINT A3 · SCREENSHOT · SAVE TO CANVA