

Sickle Cell Disease

A plain-English guide for you and your family

WHAT IS HAPPENING

Normally, red blood cells are soft, round discs filled with hemoglobin A (HbA) — the protein that grabs oxygen in the lungs and drops it off at every organ. The cells squeeze through the tiniest blood vessels (like a sponge sliding through a narrow pipe) and live about 120 days. In sickle cell disease, a child inherits two copies of a mutated gene that makes a different hemoglobin called HbS instead. When oxygen levels drop — during exercise, dehydration, cold, stress, or infection — the HbS proteins stick together like magnets, forcing the red cell to twist into a rigid crescent (sickle) shape. These stiff cells jam in small blood vessels, blocking blood flow so tissue downstream cannot get oxygen or food. The sickled cells also break apart after only 10 to 20 days, faster than the bone marrow can replace them, so the child becomes anemic (not enough red cells). The blockages cause sudden, severe pain (called a vaso-occlusive crisis) wherever blood flow stops — bones, chest, belly, or joints. Over time, repeated blockages damage the spleen (so infections become dangerous), kidneys, eyes, lungs, and brain (raising stroke risk). The body tries to cope by making red cells faster, but that cannot keep up. Treatment protects organs with hydroxyurea (which coaxes the body to make fetal hemoglobin that does not sickle), regular transfusions to dilute the sickle cells, penicillin to prevent infection, and strong pain plans for crises.

What you might feel

- Pain crises (sudden, severe pain in bones, chest, belly, or joints)
- Swelling in hands and feet (dactylitis)
- Pale skin, gums, or nail beds (anemia)
- Yellowing of the skin and eyes (Jaundice) (yellow tint to skin and whites of eyes)
- Fatigue and weakness
- Frequent infections (especially lung and bone)
- Shortness of breath
- Delayed growth and puberty
- Priapism (painful, unwanted erection lasting hours)

- Vision problems or sudden blindness

Call the doctor right away if...

- Fever over 101°F (38.3°C) — may signal a life-threatening infection because the spleen is damaged
- Sudden, severe headache or weakness on one side — could be a stroke from blocked brain blood vessels
- Chest pain, trouble breathing, or coughing — may be acute chest syndrome (sickle cells clogging lung vessels), a top cause of death
- Sudden, severe belly pain or swelling — could be splenic sequestration (the spleen traps a huge amount of blood) or liver/gallbladder blockage
- Priapism lasting more than 2–4 hours — permanent damage can happen fast

What the care team will do

- Give IV fluids at 1.5 times the maintenance rate to increase blood volume and prevent further sickling.
- Start pain medicine within 30 minutes of triage using the analgesic ladder: acetaminophen and ibuprofen first, then add opioids like morphine or hydromorphone for severe pain, and reassess pain score every 30 to 60 minutes.
- Monitor oxygen saturation continuously and give supplemental oxygen if SpO2 drops below 95% or below the patient's baseline.
- Assess respiratory rate, lung sounds, and work of breathing every 2 to 4 hours, and report new cough, chest pain, or oxygen need immediately.
- Perform incentive spirometry (breathing exercises with a device that measures deep breaths) 10 times every 2 hours while awake.
- Check temperature every 4 hours and notify the provider immediately if temperature rises above 101.3°F (38.5°C).
- Teach the patient to avoid cold exposure, dress warmly, stay hydrated with 8 to 10 glasses of water daily, and recognize early crisis symptoms.
- Verify the patient received daily penicillin prevention (prophylaxis) (preventive antibiotics) if under age 5, and administer overdue vaccinations before discharge.

Medicines you may be given — and why

(1) Hydroxyurea for medium or bad disease (makes more fetal hemoglobin, fewer crises)

(2) L-glutamine, voxelotor (raises hemoglobin), crizanlizumab (fewer crises). (3) Folic acid pills. (4) Give shots (PCV, PPSV, meningococcal, Hib, flu, COVID). (5) Penicillin pills until age 5. (6) Stop strokes — check blood vessels once a year, give transfusions if bad. (7) Bone marrow transplant or gene therapy can cure it. (8) Fix problems (chest pain, penis trouble, bone death, eye damage). (9) Make a full care plan, pain plan just for them. Each medicine on this list does a body job. Name the job before naming the drug. The medicine is the tool, the broken step is the target. Avoid the reflex of one-symptom-one-drug. Match each agent to hbS clumps when oxygen is low, makes red cells sickle, clogs blood tubes and the patients individual physiology.

Where this information comes from

- American Society of Hematology 2020 Sickle Cell Disease Guidelines
- National Heart, Lung, and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report 2014
- CDC Sickle Cell Disease Recommendations
- Cochrane Reviews on Sickle Cell Disease Interventions

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