

Sickle Cell Crisis

Sickle cell crisis happens when these bent, stiff red cells jam into tiny blood vessels like logs in a stream, blocking blood flow and starving tissue downstream—pain is just one sign of that blockage.

THE ONE-LINE TRUTH

When oxygen drops, hemoglobin S (the sickle version of the oxygen-carrying protein) turns stiff and sharp, bending red blood cells into a crescent or sickle shape.

The chain — how it unfolds

1. Vaso-occlusive pain crisis (the most common type of crisis) causes severe bone and joint pain, and in babies and toddlers it causes dactylitis (painful swelling of the hands and feet).
— why: When oxygen drops—because of dehydration, infection, cold weather, physical stress, high altitude, or even just normal sleep (when breathing slows)—hemoglobin S polymerizes and red cells sickle. Sickled cells jam into the tiny vessels that feed bones and bone marrow. Blood flow stops, and the bone tissue downstream becomes blood-starved (ischemic) (starved of oxygen). Blood-starved bone hurts terribly, and if the blockage lasts long enough, pieces of bone die (avascular tissue death (necrosis), especially in the hip and shoulder). In young children, the small bones of the hands and feet are affected first, causing dactylitis (hand-foot syndrome), often the very first sign of sickle cell disease in a baby.

2. Acute chest syndrome (ACS) causes fever, chest pain, cough, low oxygen, and a new shadow (infiltrate) on chest X-ray—it is the leading cause of death and ICU admission in sickle cell disease. — why: Sickling and vaso-blockage (occlusion) happen in the lungs just like anywhere else. Triggers include infection (pneumonia, often from *Streptococcus pneumoniae*, *Mycoplasma*, *Chlamydia*, or viruses), fat traveling clots (emboli) (tiny globs of fat released when bone marrow dies during a bone pain crisis), and in-situ sickling in pulmonary capillaries (especially if the person is already oxygen-starved (hypoxic) or dehydrated). Blocked vessels cause lung tissue to become blood-starved (ischemic), fill with fluid (pulmonary swelling (edema)), collapse (atelectasis), or get infected. The result is a vicious cycle: sickling lowers oxygen, low oxygen triggers more sickling, and the lungs fail. Without aggressive treatment—oxygen, incentive spirometry, hydration, antibiotics, transfusion—acute chest syndrome spirals into acute respiratory distress syndrome (ARDS) and can be fatal.
3. Stroke (cerebrovascular accident) can happen, especially in children; sickle cell disease is one of the most common causes of stroke in kids. — why: Sickled cells clog the large arteries at the base of the brain (especially the internal carotid and middle cerebral arteries), cutting off blood flow to brain tissue. Brain cells die within minutes without oxygen, causing sudden weakness, speech trouble, vision loss, seizures, or loss of consciousness. Chronic sickling also damages the vessel walls, causing them to narrow (stenosis) and become prone to clots. Regular transcranial Doppler (TCD) ultrasound screening in children identifies high-risk vessels, and chronic transfusion therapy (to dilute hemoglobin S and keep it below 30%) dramatically lowers stroke risk.
4. Priapism (a painful, unwanted erection lasting more than 2 to 4 hours) is a urologic emergency in males with sickle cell disease. — why: Sickled cells jam the tiny veins that normally drain blood out of the penis. Blood flows in but cannot flow out, so the penis fills with blood, becomes rigid and painful, and stays that way. The trapped, oxygen-poor blood triggers more sickling, making the blockage worse. If the blockage lasts more than 4 hours, the lack of oxygen kills the spongy erectile tissue (corpora cavernosa), causing permanent scarring, erectile dysfunction, and future episodes of priapism. Immediate treatment—hydration, pain control, sometimes aspiration of blood from the penis, and transfusion—is critical to save function.

5. Splenic sequestration crisis happens suddenly in infants and young children (usually under age 5), causing a swollen, tender belly, severe anemia, and shock. — why: In young children, the spleen is still working and full of tiny blood channels. Sickled cells get trapped in those channels and pile up, pulling a huge volume of red cells out of circulation all at once. The child's hemoglobin can drop by several grams in hours, the spleen swells (you can feel it bulging under the ribs), and blood pressure falls (hypovolemic shock). Without emergency transfusion, splenic sequestration can be fatal. After repeated episodes, or as chronic sickling damages the spleen over time, the spleen scars and shrinks (autosplenectomy), so sequestration crises stop—but the child is left without a working spleen and at high risk for life-threatening infections from encapsulated bacteria like *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Neisseria meningitidis*.
6. Aplastic crisis is a sudden, severe drop in hemoglobin caused by bone-marrow shutdown, almost always triggered by parvovirus B19 infection. — why: Parvovirus B19 (the virus that causes fifth disease, a common childhood rash illness) infects and kills red-cell precursors in the bone marrow, stopping red-cell production for about 7 to 10 days. In a healthy person, that pause is no problem—red cells live 120 days, so there are plenty in circulation to carry you through. But in sickle cell disease, red cells only live 10 to 20 days and are breaking apart constantly, so the bone marrow is already working at full speed just to keep up. When parvovirus shuts it down, hemoglobin plummets, reticulocyte count (young red cells) drops to near zero, and the person becomes dangerously anemic. Transfusion is life-saving. Once someone recovers from parvovirus B19, they are immune for life, so aplastic crisis usually happens only once.
7. Chronic destroying red blood cells (hemolytic) anemia develops over a lifetime because sickled red cells break apart (hemolyze) much faster than normal cells. — why: Day after day, sickled cells are fragile and rupture as they try to squeeze through tiny vessels or get chewed up by the spleen. The bone marrow responds by running at 6 to 8 times normal speed, pouring out new red cells as fast as it can (you will see a high reticulocyte count, large numbers of young red cells, and expanded bone-marrow spaces on imaging). But even that overdrive is not enough—steady destruction outpaces production, so baseline hemoglobin in sickle cell disease is usually 6 to 9 g/dL (compared to the normal 12 to 16). The breakdown products—free hemoglobin, heme, and bilirubin—flood the bloodstream. Free hemoglobin scavenges nitric oxide, causing blood vessels to stay tight and worsening vaso-blockage (occlusion). High bilirubin causes yellowing of the skin and eyes (jaundice) (yellow skin and eyes) and over time leads to gallstones made of bilirubin (pigment stones), often requiring surgery. The heart has to pump harder to move oxygen with fewer red cells, leading to cardiomegaly (an enlarged heart) and eventually heart failure. The spleen works overtime clearing broken cells and becomes enlarged early in life, then scars and shrinks (autosplenectomy) by age 5 to 10, leaving the person without splenic immune function and at lifelong high risk for overwhelming sepsis from encapsulated bacteria.

8. Chronic organ damage accumulates silently over years, affecting nearly every system and shortening lifespan without disease-modifying therapy. — why: Repeated cycles of vaso-blockage (occlusion), starved blood flow (ischemia), reperfusion injury, red blood cell destruction (hemolysis), and inflammation scar and destroy organs bit by bit. The kidneys lose their ability to concentrate urine (hyposthenuria), leak protein (proteinuria), and eventually fail (sickle cell nephropathy). The lungs develop pulmonary high blood pressure (hypertension) (high pressure in lung vessels) from chronic red blood cell destruction, clots, and vessel damage. The heart enlarges and weakens. The bones develop avascular tissue death (necrosis) (dead bone, especially hip and shoulder). The retina (back of the eye) loses blood flow, causing vision loss (proliferative sickle retinopathy). The skin over the ankles breaks down into chronic leg ulcers that are slow to heal. The brain suffers silent strokes (small areas of damage that do not cause obvious symptoms but add up to cognitive problems and learning difficulties). The liver becomes enlarged and scarred. Even the penis and hearing can be affected. All of this damage is driven by the same mechanism: sickling, blockage, red blood cell destruction, and inflammation, over and over, year after year.

Spot it — what you see, and why

- Severe pain in bones, especially in the back, chest, arms, or legs: Sickled-shaped red blood cells (crescent-shaped instead of round) get stuck in tiny blood vessels and block blood flow to the bone. When the bone tissue doesn't get enough oxygen, it screams with pain — like how your muscles cramp when you hold your breath during a sprint.
- Fever (temperature above 100.4°F or 38°C): Sickled cell disease damages the spleen (the body's filter that catches germs), so infections can spread fast. Fever can also signal acute chest syndrome — when sickled cells clog the lungs' blood vessels, starving lung tissue of oxygen and triggering inflammation, just like a pneumonia.
- Sudden weakness, trouble speaking, vision changes, or one-sided numbness: Sickled red blood cells can jam the arteries feeding the brain, cutting off oxygen to brain tissue — a stroke. Brain cells start dying within minutes, so every moment counts.
- Painful, unwanted erection that won't go away (priapism) in males: Sickled cells block the veins that normally drain blood out of the penis, trapping blood inside. The tissue swells and hurts intensely. If it lasts more than 4 hours, the tissue can be permanently damaged.
- Sudden shortness of breath, fast breathing, or chest pain: In acute chest syndrome, sickled cells clog lung blood vessels, fat from broken bone marrow leaks into the blood and clogs lungs, or infection inflames the lung tissue. All three cut oxygen delivery, making every breath feel like you're drowning.

- Swelling and pain in hands and feet (dactylitis), especially in babies and toddlers: Sickled cells block the small blood vessels in the bones of the fingers and toes, starving the bone and triggering swelling and throbbing pain. This is often the very first sign of sickle cell disease in young children.

Red flags — act now

- Fever above 100.4°F (38°C) — infection can turn deadly fast because the spleen is damaged
- Any sudden weakness, slurred speech, vision loss, or numbness — suspect stroke until proven otherwise
- Chest pain, trouble breathing, or coughing — may be acute chest syndrome, which can kill quickly
- Priapism (unwanted erection) lasting more than 2 hours — permanent damage starts around 4 hours
- Sudden severe belly pain or a fast-growing belly — could be splenic sequestration (spleen trapping blood) or liver crisis

Treat it — in this order

1. Intravenous (IV) fluids at 1 to 1.5 times maintenance rate using normal saline or lactated Ringer's → Hydration dilutes the blood, making it easier for sickled cells to flow through narrow vessels without getting stuck. It also replaces fluid lost from fever, fast breathing, and poor oral intake during the crisis.
2. Intravenous opioid pain medication (morphine or hydromorphone) on a scheduled basis, not just as needed, plus acetaminophen or ibuprofen if kidney function is normal → Opioids block pain signals traveling from bone tissue to the brain by attaching to opioid receptors in the spinal cord and brain. Scheduled dosing keeps pain controlled around the clock, preventing the stress and oxygen-hungry state that worsens sickling. Acetaminophen and ibuprofen reduce inflammation in the blocked tissues.
3. Supplemental oxygen by nasal cannula or mask if oxygen saturation is below the patient's baseline → Raising oxygen in the bloodstream keeps hemoglobin S from stiffening and sickling. Higher oxygen levels prevent new cells from turning into crescents and joining the blockade.
4. Incentive spirometry (a breathing device that encourages deep breaths) every 2 hours while awake, plus chest physiotherapy if needed → Deep breathing keeps the air sacs (alveoli) in the lungs inflated and moving, preventing collapse (atelectasis) and poor oxygen exchange. This reduces the risk of acute chest syndrome, which often starts when parts of the lung collapse and oxygen drops.

5. Empiric antibiotics: ceftriaxone (a third-generation cephalosporin) plus azithromycin (a macrolide) if acute chest syndrome is suspected or fever is present → Ceftriaxone kills common bacteria (like *Streptococcus pneumoniae* and *Haemophilus influenzae*) that can trigger a crisis or cause life-threatening infection in patients with sickle cell disease, whose spleens often do not work. Azithromycin covers atypical bacteria (like *Mycoplasma*) that cause acute chest syndrome.

Your top priorities at the bedside

1. Start IV normal saline at 1 to 1.5 times the maintenance fluid rate, watching lung sounds every 4 hours. — why: Hydration makes blood less thick so red cells flow better and sickle less, but giving too much fluid can cause fluid to leak into the lungs and trigger acute chest syndrome, so we give just enough to maintain good hydration without overloading.
2. Give IV opioid pain medicine on a fixed schedule every 3 to 4 hours, not waiting for the patient to ask, and add ibuprofen or ketorolac if creatinine (kidney function blood test) is normal. — why: Severe pain causes stress hormones that make more sickling happen and makes blood vessels tighten, so controlling pain breaks this cycle; scheduled dosing keeps pain controlled instead of letting it get bad between doses.
3. Apply oxygen by face mask or nasal cannula only if pulse oximetry (oxygen level monitor) shows oxygen saturation below 95 percent. — why: Low oxygen makes hemoglobin S sickle more, so we fix low oxygen right away, but giving oxygen when levels are normal can shut down the body's drive to make new red blood cells and does not help.