

MALDEK

PATIENT TEACHING · MALDEK CONDITION CATALOG

Sickle Cell Crisis

A plain-English guide for you and your family

WHAT IS HAPPENING

Normally, red blood cells are soft, round donuts that squeeze through even the tiniest blood vessels to drop off oxygen everywhere it's needed, then travel back to the lungs to reload. Hemoglobin (the protein inside each red cell that grabs and carries oxygen) picks up oxygen in the lungs and lets it go in the body's tissues, over and over. In sickle cell disease, a person inherits hemoglobin S instead of the usual type. When hemoglobin S unloads its oxygen, it can lock together into stiff rods—especially when oxygen runs low, the blood becomes more acidic, the body gets dehydrated, an infection strikes, the weather turns cold, or stress hits. Those rods poke the red cell into a crescent or sickle shape, and that sickled cell is no longer soft or round. Sickled cells pile up and block tiny blood vessels like a traffic jam. The lining of those vessels also switches on, calling in white blood cells and making everything stickier, which makes the jam even worse. Blood can't get through, so the tissue downstream starts to suffocate and die—and that is what causes the pain and damage you see in a crisis. In babies and toddlers, blocked vessels in the hands and feet cause painful swelling (dactylitis, or hand-foot syndrome). Blocked vessels in bones cause terrible bone pain, and if flow stays cut off long enough, pieces of bone die (avascular tissue death (necrosis)). When sickling and blockage happen in the lungs—often triggered by infection, fat droplets from dying bone marrow, or clumps of sickled cells—the result is acute chest syndrome: the lung fills with fluid, collapses, or gets infected, and the person struggles to breathe. If vessels in the brain get blocked, a stroke happens. If the penis vessels jam, blood gets trapped and causes priapism, a painful erection that is a medical emergency. In young children whose spleens are still working, sickled cells can suddenly flood and get stuck in the spleen (splenic sequestration), pulling so many red cells out of circulation that the child goes into shock. If parvovirus B19 (a common childhood virus) infects the bone marrow, it shuts down red-cell production for about a week; in someone with sickle cell disease—whose red cells only live 10 to 20 days instead of the normal 120—that pause triggers aplastic crisis, a sudden, severe anemia. Day in and day out, sickled cells break apart much faster than normal, so the bone marrow runs at full speed all the time trying to keep up, and the spleen (early in life) works overtime clearing the broken pieces; over the years, this chronic destruction leads to chronic destroying red blood cells (hemolytic) anemia, an enlarged heart, gallstones made of bilirubin (the yellow breakdown product of hemoglobin), and eventually a worn-out, scarred spleen that stops working. Every one of these findings traces back to the same upstream problem: hemoglobin S polymerizing (locking into rods) when oxygen is low, bending the cell, and jamming the smallest vessels. None of it is random—it is all the direct result of that one molecular change, and every complication is the body's attempt to cope with blockages and broken cells.

What you might feel

- Severe pain in bones, especially in the back, chest, arms, or legs
- Fever (temperature above 100.4°F or 38°C)
- Sudden weakness, trouble speaking, vision changes, or one-sided numbness
- Painful, unwanted erection that won't go away (priapism) in males
- Sudden shortness of breath, fast breathing, or chest pain
- Swelling and pain in hands and feet (dactylitis), especially in babies and toddlers
- Pale skin, lips, or nail beds (worse than the person's usual color)
- Yellow tint to the whites of the eyes or skin (jaundice)
- Sudden, severe belly pain (especially in the upper-right side)

Call the doctor right away if...

- 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

What the care team will do

- Start IV normal saline at 1 to 1.5 times the maintenance fluid rate, watching lung sounds every 4 hours.
- 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.
- Apply oxygen by face mask or nasal cannula only if pulse oximetry (oxygen level monitor) shows oxygen saturation below 95 percent.
- Have the patient use incentive spirometry (deep breathing device) 10 times every 2 hours while awake, and do chest physiotherapy (back patting) if ordered.
- Monitor temperature every 4 hours and report any fever of 100.4 degrees Fahrenheit or higher immediately to the provider.

- Check respiratory rate, oxygen saturation, and lung sounds every 4 hours, reporting any new cough, shortness of breath, or decreased air movement in lower lungs.
- Keep the patient warm with blankets and keep room temperature comfortable, avoiding cold drafts.
- Assess for neurologic changes every 4 hours including speech clarity, arm and leg strength, and face symmetry, using stroke scale if any concern.

Medicines you may be given — and why

Isotonic IV fluids (normal saline or lactated Ringer's)

Sickle cell crisis happens when red blood cells turn stiff and pointy because they don't have enough oxygen or water. IV fluids give water directly into the blood to keep cells from drying out and clumping together. We use maintenance rate (just enough to match what the body normally uses) because too much fluid can flood the lungs and cause acute chest syndrome.

IV opioids (morphine or hydromorphone)

Sickle cell crisis causes severe bone and muscle pain because sickled cells clog small blood vessels and starve tissue of oxygen. Opioids attach to pain receptors in the spinal cord and brain and turn down the pain signal, like lowering the volume on an alarm. We give them on a schedule (every few hours) rather than waiting for pain to spike, so the level stays steady.

NSAIDs (ketorolac or ibuprofen)

Sickled cells clog vessels and cause inflammation (swelling and chemical irritation in tissues). NSAIDs block an enzyme called cyclooxygenase (COX), which the body uses to make prostaglandins (chemicals that cause pain, fever, and swelling). Blocking COX means less prostaglandin, so less pain and inflammation. NSAIDs work well with opioids because they attack pain from a different angle.

Supplemental oxygen (nasal cannula or mask)

Sickle hemoglobin (hemoglobin S) changes shape and hardens when oxygen levels drop. Giving extra oxygen through a tube in the nose or a mask raises the oxygen level in the blood, which keeps more hemoglobin in its normal, round shape. This stops new cells from sickling. We only give oxygen if the oxygen saturation is below 95% (hypoxia), because extra oxygen when levels are already normal does not help and wastes resources.

Incentive spirometry (breathing exercise device)

Sickle cell patients in crisis often breathe shallowly because of chest or rib pain. Shallow breathing lets parts of the lungs collapse (atelectasis), which lowers oxygen and raises the risk of acute chest syndrome (a life-threatening complication). The incentive spirometer is a plastic

device that makes the patient take slow, deep breaths. Deep breaths pop open the tiny air sacs and keep lungs fully expanded.

Antibiotics: ceftriaxone (add azithromycin for acute chest syndrome)

Sickle cell patients are at high risk for serious bacterial infections because the spleen (the organ that filters bacteria from blood) is often damaged by repeated sickling. Fever in a sickle cell patient is an emergency. Ceftriaxone kills common bacteria like *Streptococcus pneumoniae* and *Salmonella*. If acute chest syndrome develops (new lung infiltrate on X-ray, chest pain, fever, cough), we add azithromycin to cover atypical bacteria like *Mycoplasma* and *Chlamydia*, which also cause pneumonia.

Red blood cell transfusion (simple or exchange)

In severe crises (stroke, acute chest syndrome not improving, multi-organ failure, or before major surgery), we replace the patient's sickled red cells with normal donor red cells. Simple transfusion adds new cells on top of the old ones. Exchange transfusion removes sickled cells and replaces them with normal cells, lowering the percentage of hemoglobin S in the blood. Normal red cells carry oxygen better, don't sickle, and flow smoothly, which can reverse life-threatening complications quickly.

Hydroxyurea

Hydroxyurea is a daily pill for long-term prevention, NOT for acute crisis. It raises the level of fetal hemoglobin (hemoglobin F) in red blood cells. Fetal hemoglobin doesn't sickle. When red cells have more hemoglobin F and less hemoglobin S, they stay round and soft even when oxygen is low. Over months, this reduces the number of crises, lowers the need for transfusions, and prevents organ damage.

L-glutamine (Endari)

L-glutamine is an amino acid (building block of protein) that red blood cells use to protect themselves from oxidative damage (injury from harmful oxygen molecules). Sick cells are under constant stress and run low on glutamine. Giving extra glutamine helps cells make antioxidants (chemicals that neutralize damage), which keeps red cells healthier and reduces sickling. It is a powder taken by mouth twice a day for long-term prevention.

Voxelotor (Oxbryta)

Voxelotor is a daily pill that binds directly to hemoglobin S inside red blood cells and stabilizes it in the oxygenated (round, soft) form. This makes it much harder for hemoglobin S to stick together and form the long, stiff chains (polymers) that cause sickling. By preventing polymerization, red cells stay flexible, live longer, and carry oxygen better. This raises hemoglobin levels over time and reduces breakdown of red cells (hemolysis).

Crizanlizumab (Adakveo)

Crizanlizumab is an IV infusion given once a month to prevent crises. It blocks a sticky protein called P-selectin on the inside of blood vessel walls and on platelets. During a crisis, P-selectin acts like Velcro, grabbing onto white blood cells and sickled red cells and making clumps that clog vessels. By covering up P-selectin, crizanlizumab stops cells from sticking, so blood flows more smoothly and crises happen less often.

Where this information comes from

- American Society of Hematology (ASH) 2020 Sickle Cell Disease Guidelines
- National Heart, Lung, and Blood Institute (NHLBI) Sickle Cell Disease Evidence-Based Management
- CDC Sickle Cell Disease Resources
- UpToDate (evidence-based clinical reference)

Last reviewed: 2026-07-07