

Acute Chest Syndrome

A plain-English guide for you and your family

WHAT IS HAPPENING

In sickle cell disease, red blood cells can change from round and flexible to stiff crescents (sickles) when oxygen drops, the blood becomes too acidic, or the body gets dehydrated. Acute chest syndrome happens when infection (like pneumonia), fat droplets from bone marrow that broke down, or sickled red blood cells clog the tiny blood vessels in the lungs. This creates a dangerous loop: blocked vessels drop oxygen, low oxygen makes more cells sickle, and more sickling blocks even more vessels. You see fever, chest pain, cough, fast breathing, and a new cloudy spot on the chest x-ray. The body tries to fix this by breathing faster to pull in more oxygen, but if the loop isn't broken with oxygen, antibiotics, blood transfusion, and breathing exercises, the lungs can fail completely. This is a medical emergency that needs hospital care right away, because it can go from stable to life-threatening in just hours.

What you might feel

- Fever (temperature 101.3°F / 38.5°C or higher)
- Sharp chest pain that gets worse with deep breathing or coughing (pleuritic chest pain)
- Cough (dry or bringing up mucus/coughed-up mucus (sputum))
- Fast breathing—more than 20 breaths per minute in adults, faster than normal for age in children (tachypnea)
- Low oxygen saturation—below 95% on room air, or a drop of 3% or more from the patient's usual level (hypoxemia)
- Shortness of breath, harder work to breathe (dyspnea)
- New cloudy or white patch on chest x-ray (pulmonary infiltrate, often in the lower parts of the lungs)
- Quieter breath sounds or crackles (popping sounds) when listening to the lungs with a stethoscope
- Coughing up blood (hemoptysis)
- Confusion or severe tiredness (altered mental status, lethargy)

Call the doctor right away if...

- Oxygen saturation below 90% even with extra oxygen given through a mask or nasal tube (signals severe lung failure coming and the lungs can't do their job anymore)
- Oxygen needs rising fast over 4 to 6 hours (suggests blocked vessels spreading through the lungs or severe whole-lung inflammation, acute respiratory distress syndrome—ARDS)
- Cloudy patches in multiple lobes (sections) of the lungs on chest x-ray (means widespread lung damage and much higher risk of death)
- Confusion, severe sleepiness, or not responding normally (signals the brain is starving for oxygen or sickled cells are blocking brain blood vessels, like a stroke)
- Coughing up blood or other signs of bleeding in the lungs (signals severe tissue death or blood vessel damage inside the lung)

What the care team will do

- Start supplemental oxygen immediately through nasal cannula at 2 to 4 liters per minute or face mask, and check oxygen saturation with a pulse oximeter every 1 to 2 hours, keeping the level above 95%
- Give intravenous fluids at 1 to 1.5 times the normal daily amount (maintenance rate) using normal saline or D5 half-normal saline, but watch closely for fluid overload—check lung sounds every 4 hours and stop fluids if you hear crackles or the patient gains more than 1 kilogram in 24 hours
- Administer intravenous broad-spectrum antibiotics within 1 hour of diagnosis—ceftriaxone 1 to 2 grams IV once daily PLUS azithromycin 500 milligrams IV once daily—and document the exact time given
- Coach the patient to use an incentive spirometer (a handheld device with a ball that rises when you breathe in) every 2 hours while awake, taking 10 slow deep breaths each time, and record the volume reached
- Give strong pain medicine—usually intravenous morphine 0.1 milligrams per kilogram every 2 to 4 hours or a continuous morphine drip, or hydromorphone for severe pain—and use a pain scale every 2 hours, keeping pain at 3 out of 10 or lower
- Monitor respiratory rate, oxygen saturation, work of breathing (look for flared nostrils, using neck or belly muscles to breathe, or ribs sucking in), and mental status every 2 to 4 hours, and call the provider immediately if oxygen saturation drops below 92%, respiratory rate goes above 24, or the patient becomes confused

- Notify the hematology team (blood specialists) within 1 hour of diagnosis and prepare for possible simple transfusion (giving 1 to 2 units of packed red blood cells) or exchange transfusion (removing the patient's sickled blood and replacing it with normal donor blood) if oxygen needs increase or chest x-ray shows worsening cloudy areas
- Keep the head of the bed elevated at 30 to 45 degrees unless the patient has low blood pressure, and help the patient turn from side to side every 2 hours

Medicines you may be given — and why

Oxygen (supplemental)

Oxygen is a gas you breathe in through a mask or nasal prongs. It raises the amount of oxygen dissolved in your blood. When red blood cells are bathed in high oxygen, hemoglobin S (the sickle type) is less likely to clump together and twist the cell into a rigid crescent shape. This reduces new sickling and helps keep small blood vessels open so blood can flow.

Ceftriaxone

Ceftriaxone is a bacteria-killing drug. It works by breaking the outer wall that bacteria need to survive. Without that wall, bacteria burst and die. We use it in acute chest syndrome because infection (pneumonia) is a common trigger, and ceftriaxone kills the most likely germs—*Streptococcus pneumoniae*, *Haemophilus influenzae*, and some others.

Azithromycin

Azithromycin stops bacteria from making the proteins they need to grow and multiply. It locks onto the bacteria's protein-building factory (the ribosome) so the factory cannot read its instructions. The bacteria cannot make new parts, so they stop growing and eventually die. We add azithromycin to ceftriaxone because it kills atypical bacteria (*Mycoplasma pneumoniae* and *Chlamydia pneumoniae*) that ceftriaxone misses.

Morphine or Hydromorphone

These drugs block pain signals in your brain and spinal cord. They attach to opioid receptors (like a key fitting a lock) and tell the nervous system to turn down the pain volume. In acute chest syndrome, strong pain control is critical—when chest pain is relieved, you can take deep breaths and cough to clear mucus, which keeps airways open and prevents more lung collapse.

Intravenous fluids (normal saline or balanced crystalloid)

IV fluids are sterile salt water given through a vein. They keep your blood volume up and your blood flowing smoothly. Dehydration (too little water in the body) makes blood thicker and sickle cells stickier, which worsens vessel blockage. The goal is euvolemia—just the right

amount of fluid, not too little (which thickens blood) and not too much (which can flood the lungs).

Incentive spirometer (breathing device)

This is a handheld device with a mouthpiece and a tube with a floating ball or piston. You breathe in slowly and deeply, which sucks the ball up. It trains you to fully inflate your lungs, especially the lower parts (bases) that tend to collapse when you breathe shallowly from pain. Deep breaths open tiny airways (alveoli) and prevent atelectasis (collapsed lung areas), which would worsen oxygen levels and trigger more sickling.

Simple red blood cell transfusion

This is an infusion of donated red blood cells (from a healthy person) into your vein. The new cells carry hemoglobin A (normal, flexible hemoglobin) instead of hemoglobin S. They raise your total hemoglobin level, improve oxygen delivery to tissues, and dilute the percentage of sickled cells. We use simple transfusion when hemoglobin drops below baseline (usually to 5–7 g/dL or lower) or if oxygen delivery is critically low.

Red blood cell exchange transfusion (erythrocytapheresis)

Exchange transfusion removes your blood (high in sickled cells) and replaces it with healthy donor blood at the same time. A machine takes blood out of one vein, separates and discards your red cells, and returns donor red cells plus your own plasma and platelets through another vein. The goal is to drop your hemoglobin S percentage below 30% quickly. We use exchange in severe acute chest syndrome (multi-lobe pneumonia, very low oxygen, or rapid worsening) because it works faster and more completely than simple transfusion.

Bronchodilators (e.g., albuterol inhaler or nebulizer)

Albuterol relaxes the smooth muscle wrapped around your airways (bronchioles). When those muscles tighten (bronchoconstriction), airways narrow and air cannot flow well—you wheeze and feel short of breath. Albuterol tells the muscle to relax by activating beta-2 receptors, which makes the muscle cells loosen their grip. This opens the airways wider so you can breathe easier. We use it in acute chest syndrome if there is wheezing or a history of asthma.

Where this information comes from

- American Society of Hematology 2020 Guidelines for Sickle Cell Disease
- National Heart, Lung, and Blood Institute (NHLBI) 2014 Evidence-Based Management of Sickle Cell Disease
- American Thoracic Society
- JAMA 2000 National Acute Chest Syndrome Study Group

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