CONDITION

# Sickle cell crisis

A sickle cell crisis is an acute episode of pain or illness caused by sickled red blood cells.

**WHAT IT IS**
Acute pain or illness caused by sickled blood cells

**BODY SYSTEM**
Blood and circulation

**MAIN SYMPTOMS**
Severe pain, swelling, fever, breathing difficulty

**WHAT CAUSES IT**
Inherited sickle cell disease, sometimes with triggers

**HOW IT IS DIAGNOSED**
Clinical assessment and tests for complications

**MAIN TREATMENT**
Rapid pain relief, supportive care, and complication-specific treatment

**CAN IT BE CURED?**
Sometimes - specialist therapies can cure underlying sickle cell disease

**IS IT AN EMERGENCY?**
Sometimes - with fever, chest symptoms, stroke signs, or uncontrolled pain

**LONG-TERM OUTLOOK**
Episodes may recur, but preventive treatment can reduce their impact

## Overview

A sickle cell crisis is a sudden episode related to sickle cell disease. Most often, the term means a vaso-occlusive pain crisis . This happens when unusually shaped red blood cells restrict blood flow through small blood vessels, causing pain and inflammation.

Sickle cell disease is an inherited blood condition. It affects hemoglobin, the protein in red blood cells that carries oxygen. Under certain conditions, affected cells can become stiff and curved like a sickle. These cells may break down early or block blood flow.

A crisis can affect almost any part of the body. Pain commonly occurs in the back, chest, arms, legs, abdomen, or joints. Some episodes can be managed using a personal home-care plan. Others require urgent assessment or hospital treatment.

The word "crisis" can also describe complications other than pain, including acute chest

syndrome, sudden severe anemia, or trapping of blood in the spleen. These complications need prompt medical care.

## Symptoms

Pain from a vaso-occlusive crisis may begin suddenly or build over several hours. It can feel sharp, throbbing, aching, or deep. The location and intensity may change during an episode.

Possible symptoms include:

Not every new symptom is caused by a pain crisis. For example, chest pain and breathing difficulty may indicate acute chest syndrome , a serious lung complication. Sudden weakness, facial drooping, speech changes, or loss of balance may be signs of a stroke.

Pain can also become chronic, meaning it is present on most days or continues between acute episodes. Chronic pain and an acute crisis can occur at the same time.

- Moderate to severe pain, often in the bones, joints, chest, back, or abdomen
- Swelling of the hands or feet, especially in young children
- Tiredness, weakness, or reduced activity
- Yellowing of the eyes or skin from red blood cell breakdown
- Fever or other signs of infection
- Shortness of breath, coughing, or chest pain
- Headache, dizziness, confusion, or weakness

## Causes and risk factors

Sickle cell crisis occurs in people with sickle cell disease. The disease is caused by inherited changes in the gene that provides instructions for making hemoglobin. It is not caused by lifestyle, and it cannot be caught from another person.

A crisis begins when sickled cells, inflammation, and changes in blood vessels reduce blood flow to tissues. The exact reason an individual episode starts is not always clear.

Possible triggers include:

A crisis can occur even when you have avoided known triggers. Having an episode does not mean that you did something wrong.

The frequency and severity of crises differ among people. Sickle cell type, previous

complications, access to preventive treatment, pregnancy, and other medical conditions can all affect risk.

- Dehydration
- Infection or fever
- Sudden temperature changes
- Very cold conditions
- Low oxygen levels, including at high altitude
- Intense physical exertion without enough rest or fluids
- Emotional stress
- Poor sleep
- Surgery or another major illness

## Diagnosis

If you already have confirmed sickle cell disease, a pain crisis is usually identified from your symptoms, medical history, and physical examination. There is no single blood test that proves pain is caused by blocked blood flow.

Tests may be used to look for anemia, infection, organ problems, or other causes of pain. These may include a complete blood count, a count of newly produced red blood cells, kidney and liver tests, and checks of oxygen levels.

Chest pain, coughing, fever, or breathing difficulty may require a chest X-ray and other tests for acute chest syndrome or infection. Neurological symptoms may require urgent brain imaging. Blood cultures may be taken when a serious bacterial infection is possible.

If sickle cell disease has not previously been diagnosed, specialized hemoglobin testing can identify abnormal hemoglobin. Tests include hemoglobin electrophoresis or high-performance liquid chromatography. Genetic testing is sometimes used to clarify the type.

## Treatment and management

Treatment depends on the symptoms, their severity, and whether a complication is present. A personal pain plan can help you and the healthcare team act quickly.

Pain relief is an early priority. Mild, familiar pain may sometimes be managed at home using

prescribed or recommended medicines, fluids, warmth, rest, and gentle movement. Anti-inflammatory medicines may not be suitable for everyone, particularly people with kidney, stomach, or bleeding problems.

Moderate or severe pain may require opioids and close monitoring in an emergency department or hospital. Treatment should take account of medicines that have helped before. Needing opioids like oxycodone or morphine for severe sickle cell pain is not, by itself, evidence of addiction.

Fluids may be given if you are dehydrated or cannot drink. Excess fluid can be harmful, so intravenous fluids are not automatically needed in every crisis. Oxygen is generally used when the oxygen level is low rather than as routine pain treatment.

Other treatment may include:

A straightforward pain crisis does not routinely require a blood transfusion.

Long-term treatments can reduce future episodes. Hydroxyurea helps the body produce more fetal hemoglobin, a form of hemoglobin that is less likely to sickle. Other disease-modifying medicines may be appropriate for some people. These treatments require specialist assessment and monitoring.

Stem cell transplantation can cure sickle cell disease in some people. Gene-based treatments are also available for selected patients in some healthcare systems. These options involve major treatment, significant risks, and detailed specialist assessment.

- Antibiotics like amoxicillin or doxycycline for suspected or confirmed bacterial infection
- Breathing exercises using an incentive spirometer, especially with chest or upper-body pain
- Blood transfusion for selected complications, such as severe anemia, stroke, or acute chest syndrome
- Treatment for the specific cause of pain when it is not a typical crisis

## Complications

A crisis can sometimes lead to serious complications. These are more than an increase in pain and often cause additional symptoms.

Acute chest syndrome causes a new lung problem with symptoms such as chest pain, fever, coughing, low oxygen, or trouble breathing. It can worsen quickly and usually requires hospital treatment.

Stroke can occur when blood flow in the brain is blocked or when a blood vessel bleeds.

Sudden facial drooping, one-sided weakness, speech difficulty, confusion, severe headache, or loss of coordination requires emergency help.

Severe anemia occurs when too few red blood cells remain to carry enough oxygen. It may follow increased cell breakdown, temporary slowing of blood cell production, or rapid trapping of blood in the spleen. Symptoms can include extreme tiredness, pale skin, faintness, a rapid heartbeat, or shortness of breath.

Other complications include serious infection, kidney injury, gallstones, bone or joint damage, leg ulcers, and a prolonged painful erection called priapism. Repeated crises may contribute to long-term organ damage, although regular monitoring and preventive treatment can reduce some risks.

## Prevention

Sickle cell disease is inherited and cannot be prevented through diet, exercise, or other lifestyle changes. Not every crisis can be prevented.

Disease-modifying treatment can reduce pain episodes and other complications for many people. Taking prescribed treatment consistently and attending routine sickle cell appointments are important parts of prevention.

Practical measures may also lower the chance of some episodes:

These steps reduce risks but cannot guarantee that a crisis will not happen.

- Drink regularly, especially during hot weather, exercise, or illness
- Avoid becoming extremely cold or overheated
- Build up physical activity gradually and take rest breaks
- Follow the recommended vaccination and infection-prevention plan
- Avoid smoking and second-hand smoke
- Prepare for travel to high altitudes or places with limited medical care
- Keep a written pain and emergency plan available

## Living with it and what to expect

A pain crisis may last for several hours or several days. Recovery can continue after the worst pain has passed, with tiredness, poor sleep, stiffness, or difficulty concentrating. Some episodes last longer or return soon after treatment.

The pattern can change over months and years. Some people have long periods between crises, while others experience frequent episodes or chronic pain. Previous history can offer clues, but it cannot predict every future crisis.

Sickle cell disease is lifelong unless successfully treated with a curative therapy. Regular follow-up usually includes checks for anemia and damage involving the kidneys, lungs, eyes, bones, and other organs. Preventive treatment may change over time as your health and circumstances change.

Crises can disrupt school, employment, sleep, family life, and mental health. Planning may include flexible work or school arrangements, access to pain treatment, and support for anxiety, low mood, or trauma linked to repeated hospital care.

Keeping a record of symptoms, possible triggers, medicines, and what helped can make future treatment more consistent. A clear care plan is especially useful when you are seen by a team that does not know you.

## When to get medical help

Contact your usual sickle cell team promptly if episodes are becoming more frequent, pain is changing, home treatment is less effective, or medicine side effects are causing problems. These concerns usually need timely follow-up but are not automatically emergencies.

Seek urgent medical assessment for:

Call emergency services for trouble breathing, severe chest pain, blue or grey lips, fainting, a seizure, unusual difficulty waking, or signs of stroke. Stroke signs include facial drooping, weakness on one side, speech difficulty, sudden confusion, loss of balance, or a sudden severe headache.

A painful erection lasting four hours is also an emergency because delayed treatment can cause lasting damage.

- Pain that is severe, unusual, or not improving with your home plan
- Fever, especially 38.5°C or higher, or the threshold in your care plan
- Repeated vomiting or difficulty drinking
- Marked weakness, paleness, or a racing heartbeat
- A painful erection that persists or keeps returning
- New abdominal swelling, particularly in a child

## Common questions

How long does a sickle cell crisis last?

Many pain crises last several hours to several days. Some continue longer, and tiredness or soreness may remain after the strongest pain settles. Seek medical help if pain is unusually severe, different from normal, or not responding to your care plan.

What triggers a sickle cell crisis?

Common triggers include dehydration, infection, temperature changes, low oxygen, intense exertion, and stress. However, many crises have no clear trigger. An unexplained episode is not evidence that you failed to prevent it.

Can I treat a sickle cell crisis at home?

A mild, familiar episode may sometimes be managed at home using your agreed pain plan, fluids, warmth, and rest. Get urgent help if the pain is not improving or you develop fever, chest symptoms, breathing difficulty, unusual weakness, or neurological symptoms.

Do I need to go to hospital for every crisis?

Not necessarily. This depends on the severity of pain, your response to home treatment, and whether warning signs are present. Chest symptoms, stroke signs, severe anemia symptoms, fever, or uncontrolled pain need prompt medical assessment.

Can sickle cell trait cause a pain crisis?

Sickle cell trait is not the same as sickle cell disease and does not usually cause typical pain crises. Rare complications can occur under extreme conditions, such as severe dehydration, very low oxygen, or intense exertion at altitude. New or severe pain still needs an appropriate medical assessment.

Can sickle cell crises stop permanently?

Preventive medicines can greatly reduce crises for some people but do not guarantee that they will stop. Stem cell transplantation or gene-based treatment can cure the underlying disease in selected people, but these treatments are not suitable or available for everyone.

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