



Sickle cell anemia is a chronic hemolytic anemia. It is characterized by the presence of sickle-shaped red blood cells. Sickle cell crisis results from the occlusion of a blood vessel by masses of these misshapen blood cells. Pain is the principal manifestation and represents the most common type of crisis. Typical pain occurs in the joints and back. Hepatic, pulmonary, or central nervous system involvement can occur, with each type being associated with its own group of symptoms. Keep in mind that patients with sickle cell disorder have a high incidence of life-threatening disorders at a very young age.

A. Sickle Cell Crisis

A sickle cell crisis is a painful episode that occurs in people who have sickle cell anemia. It happens when sickle-shaped red blood cells (RBCs) block blood vessels. Blood and oxygen cannot get to the tissues, causing pain. Also, a sickle cell crisis can damage your tissues, cause organ failure and can potentially become life-threatening.

Adult Care

EMR/BLS

1. Initial Assessment/Care [Protocol 1.](#)
2. Administer 100% oxygen via non-rebreather mask at 15L/min.
3. Provide emotional support, calm and reassure.
4. Apply hot packs to affected areas, especially joints and areas of increased pain.

ALS

5. Establish IV access of Normal Saline.
6. Administer **Normal Saline up to 1000 mL** assess B/P and lung sounds often.
7. If pain persists and systolic BP > 90 mmHg:
 - a) Administer **Fentanyl 50 mcg** slow IVP. May be repeated once, the second dose can be titrated up to 50 mcg until relief of pain relief (Max total dose of 100 mcg) [Medication 16.](#)
 - b) If IV/IO is not available, may administer Fentanyl Intranasal 50 mcg (max 1 mL/nostril). May repeat dose once 50 mcg (1 mL/nostril).
 - c) Administer **Zofran 4 mg** SL (Do not administer to pregnant females) [Medication 33.](#)



B. Transport

Patterns of an acute sickle cell crisis are now recognizable. They are based on the part of the body where the crisis occurs.

- Acute chest syndrome: Sudden acute [chest pain](#) with [coughing](#) up of blood can occur. Low-grade fevers can be present. The person is usually short of breath. If a [cough](#) is present, it often is nonproductive. Acute chest syndrome is common in a young person with sickle cell disease. Chronic (long-term) sickle cell lung disease develops over time because the acute and subacute lung crisis leads to scarred lungs, as well as, other problems.
- Abdominal crisis: The pain associated with the abdominal crisis of sickle cell disease is constant and sudden. It becomes unrelenting. The pain may or may not be localized to any one area of the abdomen. [Nausea](#), [vomiting](#), and [diarrhea](#) may or may not occur.
- Joint crisis: Acute and painful joint crisis may develop without a significant traumatic history. Its focus is either in a single joint or in multiple joints. Often the connecting bony parts of the joint are painful. Range of motion is often restricted because of the pain. [Avascular necrosis](#) of the hips can occur, causing permanent damage.

Any patient with a history of sickle cell anemia presenting with any of the above three syndromes will be treated/transported ALS to the closest appropriate facility.