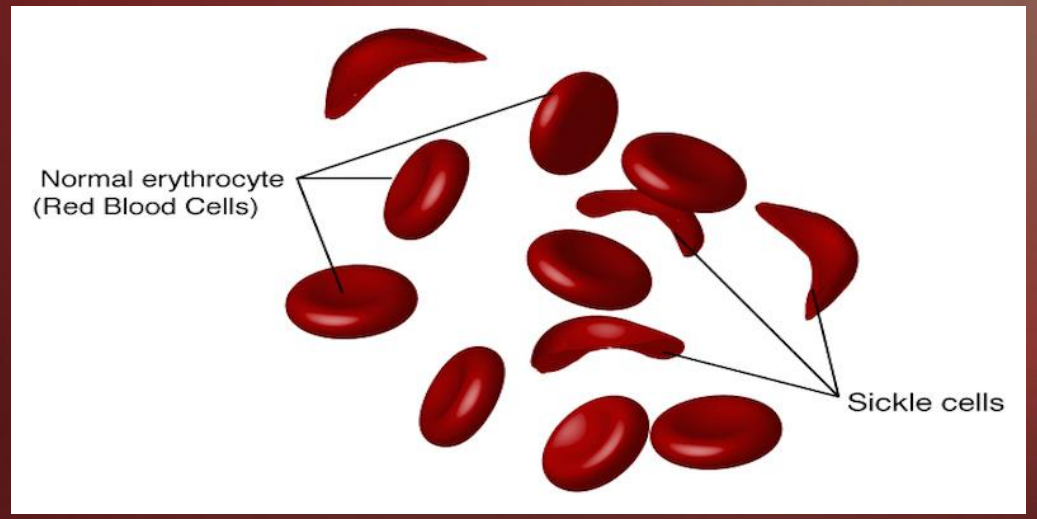
SICKLE CELL DISEASE: PRACTICE GUIDELINES

WHAT IS SICKLE CELL DISEASE?

Sickle cell disease is an inherited life-threatening blood disorder that affects the red blood cells, causing them to sickle. These sickled cells carry less oxygen throughout the body and can clump together in small vessels, blocking off blood supply, and causing organ damage as well as pain.



TYPES OF SICKLE CELL

There many different forms of sickle cell disease. The most common types are:

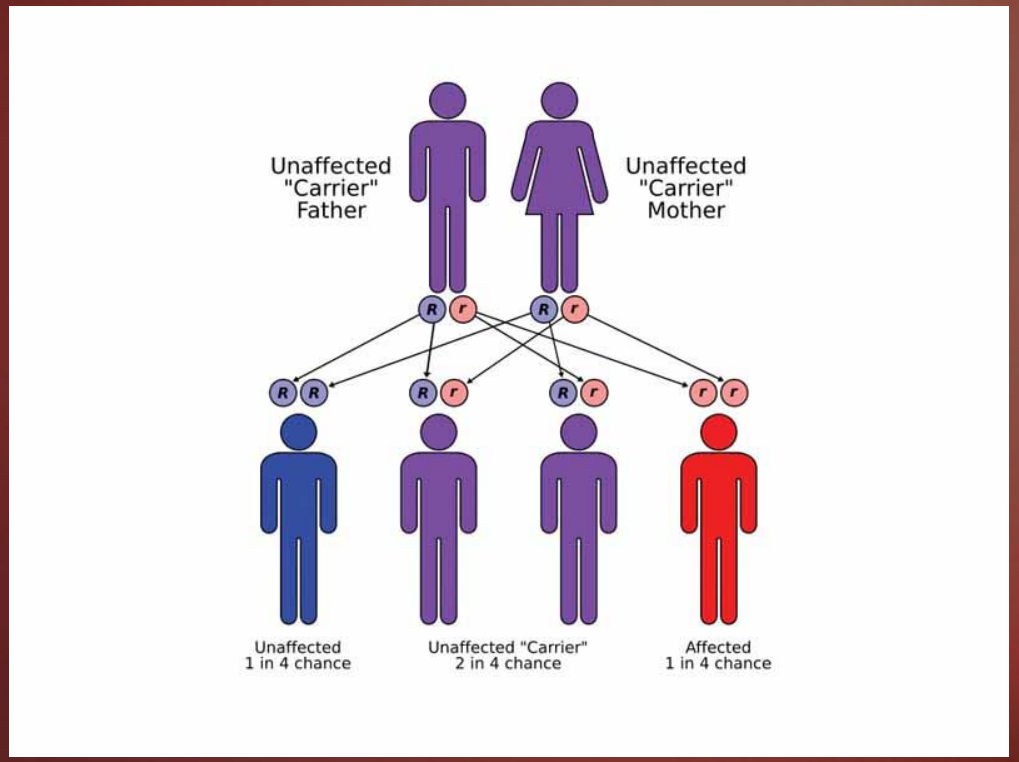
- **Hemoglobin SS disease**
- **Hemoglobin SC disease**
- **Hemoglobin SB+ (beta) thalassemia**
- **Hemoglobin SB 0 (Beta-zero) thalassemia**

HOW IS SICKLE CELL INHERITED?

Sickle cell disease is inherited when both biological parents carry a form of the recessive trait.

Carriers do not typically show signs or symptoms of sickle cell, except under extreme conditions.

Every child that is conceived by two carriers has a 2 in 4 chance of carrying the trait and 1 in 4 chance of inheriting sickle cell disease.

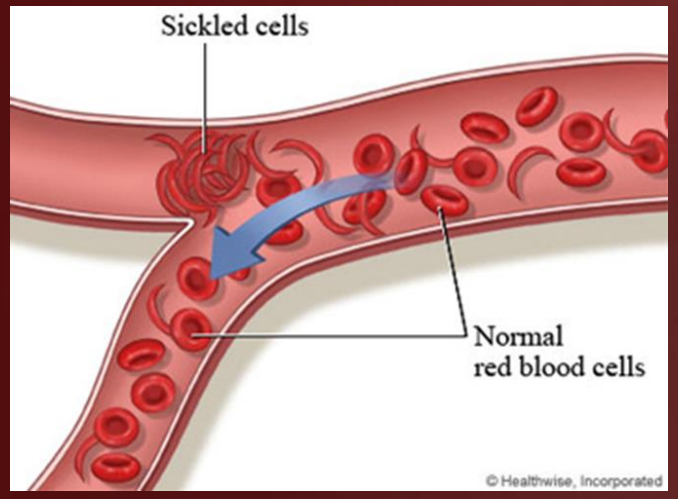


COMPLICATIONS OF SICKLE CELL DISEASE

- Hand-Foot Syndrome
- Pain
- Anemia
- Infection
- Acute Chest Syndrome
- Splenic Sequestration
- Vision Loss
- Leg Ulcers
- DVT and PE
- Stroke
- Organ Damage
- Malnutrition/Growth Retardation
- Gallstones
- Priapism

SICKLE CELL PAIN

Pain is a common symptom of sickle cell disease. These episodes of pain, called crises, may vary in location, intensity, and duration. Some episodes may last a few hours, while others may last a few weeks or more. The frequency of pain episodes also vary from person to person. Some children experience chronic pain as well, which can result from bone and joint damage.



CORRELATION OF ASTHMA AND SICKLE CELL

Many children with sickle cell disease will also have asthma-like symptoms. This is often due to damage that has occurred in the lungs from the disease, but may also be due to asthma itself.

Most children will be seen by a Hematologist-Pulmonologist team. They work together to keep the patient's lungs and airways open by the use of long-term and emergency inhalers. By allowing better oxygenation to the blood, sickling episodes are prevented or made less severe.

SICKLE CELL PAIN TRIGGERS

- Exposure to cold/hot temperatures
- Dehydration
- Exercise
- Fever
- Smoking
- Bacterial infection:
  - Strep throat
  - UTI
  - Pneumonia
- Viral illness:
  - Influenza
  - Viral exanthem (rash)

THE IMPORTANCE OF HYDRATION

Hydration is a key factor in preventing sickle cell pain as well as many other symptoms. Being hydrated helps keep the sickled cells from clumping together and getting stuck in small blood vessels.

Staying properly hydrated can be a challenge for those with the disease. Kidney damage due to sickle cell causes the inability of the kidneys to concentrate urine. The kidneys in turn dump excessive amounts of fluid.

Sweating in hot weather or during exercise and drinking caffeinated beverages can also cause sickle cell symptoms.

SUGGESTED DAILY FLUID INTAKE

Body Weight	Number of 8oz. Glasses of water
30 pounds	5-8
45 pounds	6-9
55 pounds	7-10
75 pounds	8-11
100 pounds	9-13
130 pounds	10-15
150 pounds	11-17
175 pounds	12-18

WHAT TO DO DURING A PAIN EPISODE

- Have the child relax and provide them water to drink
- Administer pain medication, try to start by giving Ibuprofen first, unless care plan states otherwise
- Put a heating pad on the affected area
- Communicate with parent and child on what works best for them to get relief
- DO NOT use ice on anyone with sickle cell disease.
- Encourage deep breathing and use portable oxygen if available

COMMON ACUTE COMPLICATIONS

- Acute chest syndrome
- Fever
- Hepatobiliary Complications
- Multisystem organ facilure
- Pain from a vaso-occlusive crisis
- Priapism
- Splenic Sequestration
- Stroke

TREATMENT AND MANAGEMENT

*Following recommendations are as detailed in the American Society of Hematology guide for clinicians

*These recommendations are adapted from the National Heart, Lung, and Blood Institute's *Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014*

ACUTE CHEST SYNDROME

- Resembles pneumonia
- Increased risk associated with individuals with asthma or prior ACS events
- Most common cause of death in adults with SCD
- Evaluation: Should include chest x-ray, pulse oximetry, CBC/reticulocyte count, blood culture
- Treatment: Intravenous cephalosporin and oral macrolide antibiotic; supplemental oxygen, incentive spirometry

Rapid progress of ACS there is a clinical recommendation for blood exchange

PAIN FROM VASO-OCCLUSIVE CRISIS

- "There is no test for VOC, only tests to rule out other causes of pain, eg, avascular necrosis, cortical bone infarction, osteomyelitis, acute chest syndrome, infection, or other abdominal complications"

Patient self report is vital! Most SCD patients know what works for them to control their pain. Please take into account history of opiate use, possible tolerance, collateral reports from family, friends.

TREATMENT GUIDELINES

Begin to treat acute pain while evaluating other causes

Rapidly initiate analgesic therapy within 30 minutes of triage, 90 minutes of registration

Base analgesic selection on pain assessment, associated symptoms, outpatient analgesic use, patient knowledge of effective agents and does and past experiences with side effects

TREATMENT GUIDELINES CONTINUED

- For mild to moderate pain, in patients who report relief with NSAIDs, and absent contraindications, continue treatment with NSAIDs
- For severe pain, use parenteral opioid
- Encourage incentive spirometry while awake, and ambulation as soon as possible
- Use adjunctive nonpharmacological approaches to treat pain such as local heat application, distraction
- Oxygen, IV fluids if clinically indicated

OPIOID GUIDELINES FOR SEVERE, ACUTE PAIN

- Keep it individualized!
- Do not use meperidine (Demerol) unless it is the only effective opioid for an individual patient
- Calculate the parenteral dose based on total home dose
- Administer meds subcutaneous route if intravenous route is difficult
- Reassess pain and re-administer opioids if necessary for continued pain (every 15-30 minutes) until pain is under control (per patient report)

OPIOID GUIDELINES CONTINUED

- Maintain or consider escalation of dose by 25 percent until pain is controlled
- Reassess after each dose for pain relief and side effects
- Monitor for excessive sedation by using sedation measurement scale and by obtaining oxygenation levels
- Gradually titrate down parenteral opioids as pain resolves
- In discharge planning, wean parenteral opioids prior to conversion to oral opioids (adjust as needed) to prevent withdrawal

TO LEARN MORE

- Please visit our website www.sicklecelltx.org
- Call us at 1-844-994-HOPE
- Visit American Society of Hematology for practice guidelines