

LIVING WITH:

# SICKLE CELL DISEASE

A GUIDE TO DAILY LIVING

## TABLE OF CONTENTS

**03**

Managing The Pain

**05**

Treating Complications

**06**

Lifestyle Changes For  
Sickle Cell

**08**

Stay Fit With Sickle  
Cell

**11**

Finding The Support  
You Need

**13**

Specialists to Have

**15**

Emergency Room Tips

<b>03</b>	Managing The Pain
<b>05</b>	Treating Complications
<b>06</b>	Lifestyle Changes
<b>08</b>	Stay Fit
<b>11</b>	Find Support

# Sickle Cell Disease in the United States

- SCD affects approximately 100,000 Americans.
- SCD occurs among about 1 out of every 365 Black or African-American births.
- SCD occurs among about 1 out of every 16,300 Hispanic-American births.
- About 1 in 13 Black or African-American babies is born with sickle cell trait (SCT).



# Managing The Pain

Because there is no guaranteed cure, treatment for sickle cell anemia is usually aimed at avoiding crises, relieving symptoms and preventing complications.

If you have sickle cell anemia, you'll need to make regular visits to your doctor to check your red blood cell count and monitor your health.

Treatments may include medications to reduce pain and prevent complications, blood transfusions and supplemental oxygen, as well as bone marrow transplant.

## **Antibiotics:**

Children with sickle cell anemia usually begin taking the antibiotic penicillin when they're about 2 months of age and continue taking it until they're 5 years old.

Doing so helps prevent infections, such as pneumonia, which can be life-threatening to an infant or child with sickle cell anemia. Antibiotics may also help adults with sickle cell anemia fight certain infections.

## **Pain Relieving Medication:**

To relieve pain during a sickle crisis, your doctor may advise over-the-counter pain relievers and application of heat to the affected area. You may also need stronger prescription pain medication.

## **Supplemental Oxygen:**

Breathing supplemental oxygen through a breathing mask adds oxygen to your blood and helps you breathe easier. It may be helpful if you have acute chest syndrome or a sickle cell crisis.





# Managing The Pain

## **Hydroxyurea (Droxia, Hydrea):**

This prescription drug, normally used to treat cancer, may be helpful for adults with severe disease.

When taken daily, it reduces the frequency of painful crises and may reduce the need for blood transfusions. It seems to work by stimulating the production of fetal hemoglobin – a type of hemoglobin found in newborns that helps prevent the formation of sickle cells.

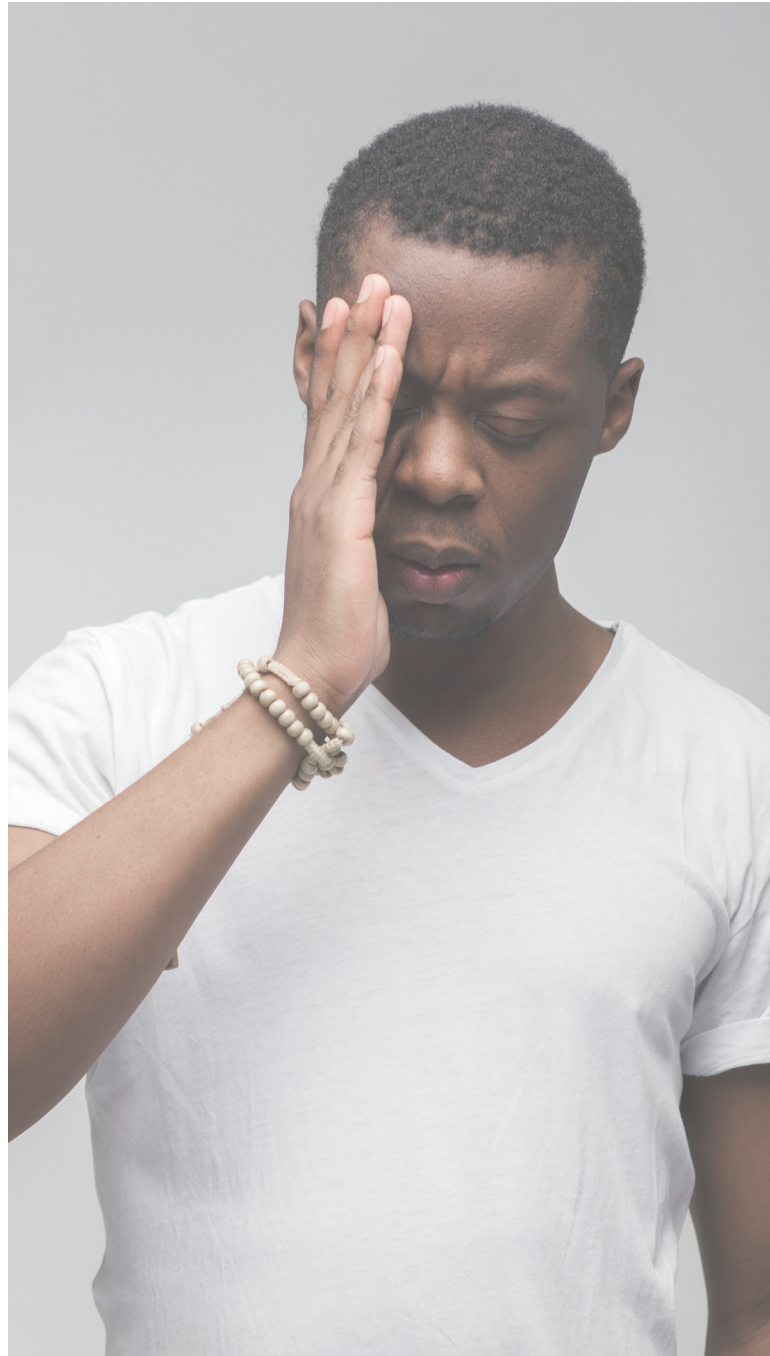
There is some concern about the possibility that long-term use of this drug may cause tumors or leukemia in certain people. Your doctor can help you determine if this drug may be beneficial for you.

## **Bone Marrow Transplant:**

This procedure replaces bone marrow affected by sickle cell anemia with healthy bone marrow from a donor who doesn't have the disease. It can be a cure, but the procedure is risky, and it's difficult to find suitable donors. Researchers are still studying bone marrow transplants for people with sickle cell anemia.

Currently, the procedure is recommended only for people who have significant symptoms and problems from sickle cell anemia.

Bone marrow transplant requires a lengthy hospital stay. After the transplant, you'll need drugs to help prevent rejection of the donated marrow.



# Treating Complications

## **Treating Complications:**

Doctors treat most complications of sickle cell anemia as they occur. Treatment may include antibiotics, vitamins, blood transfusions, pain-relieving medicines, other medications and possibly surgery, such as to correct vision problems or to remove a damaged spleen.

### **Butyric Acid**

Normally used as a food additive, butyric acid may increase the amount of fetal hemoglobin in the blood.

### **Clotrimazole**

This over-the-counter antifungal medication helps prevent a loss of water from red blood cells, which may reduce the number of sickle cells that form.

### **Nicosan**

This is an herbal treatment in early trials in the U.S. Nicosan has been used to prevent sickle crises in Nigeria.

## **Gene Therapy**

Because sickle cell anemia is caused by a defective gene, researchers are exploring whether inserting a normal gene into the bone marrow of people with sickle cell anemia will result in the production of normal hemoglobin.

Scientists are also exploring the possibility of turning off the defective gene while reactivating another gene responsible for the production of fetal hemoglobin – a type of hemoglobin found in newborns that prevents sickle cells from forming.

### **Nitric Oxide**

Sickle cell anemia causes low levels of nitric oxide, a gas that helps keep blood vessels open and reduces the stickiness of red blood cells. Treatment with nitric oxide may prevent sickle cells from clumping together.





# Make The Change

Now's a great time to create lifestyle habits that will do their part in helping control sickle cell pain, maintain your health and prevent crises.

Remember that infants and children with sickle cell disease need to receive regular childhood vaccinations. Children and adults with sickle cell anemia also should have a yearly flu shot and be immunized against pneumonia. If you or your child has sickle cell anemia, follow these additional suggestions to help stay healthy.

## **Diet & Vitamins**

Take folic acid supplements daily and eat a balanced diet. Bone marrow needs folic acid and other vitamins to make new red blood cells.

## **Hydration Nation**

Drink plenty of water. Staying hydrated helps keep your blood diluted, which reduces the chance that sickle cells will form.

## **Keep It Steady**

Avoid temperature extremes. Exposure to extreme heat or cold can trigger the formation of sickle cells.

## **Move Your Body**

Exercise regularly, but don't overdo it. Talk to your doctor about how much exercise is right for you.

## **Don't Trip**

Reduce stress. A sickle crisis can occur as a result of stress.

# Make The Change (continued)

## Use With Caution

Use over-the-counter medications with caution. Some medications, such as the decongestant pseudoephedrine, can constrict your blood vessels and make it harder for the sickle cells to move through freely.

## Stay Low

Avoid high-altitude areas as much as possible. Traveling to a high-altitude area may also trigger a crisis because of lower oxygen levels

## Prepare To Take Flight

Fly on airplanes with pressurized cabins. Unpressurized aircraft cabins may not provide enough oxygen.

Low oxygen levels can trigger a sickle crisis. Additionally, be sure to drink extra water when traveling by air, as pressurized cabins can be dehydrating.



*Change your habits to change your life.*





# STAY FIT WITH SICKLE CELL

**CHRONIC PAIN IS ONE OF THE MOST COMMON COMPLICATIONS OF SICKLE CELL DISEASE (SCD). AVOIDING ACTIVITIES THAT CAN TRIGGER A FULL BLOWN PAIN "EPISODE" OR "CRISIS" IS KEY TO STAYING HEALTHY AND OUT OF THE ER. FOR SOME, THIS INCLUDES AVOIDING EXERCISE... BUT, IT DOESN'T HAVE TO.**



**Don't Let Sickle Cell Keep You Down**

Strenuous exercise, like taking a boot camp class or hiking, reduces oxygen levels in the blood and may cause red blood cells to sickle. The cells become hard and sticky C shapes and as they travel through tiny blood vessels, they can become stuck and painfully clog blood flow. Low - to moderate-impact exercise, on the other hand, can help to increase oxygen, keep blood circulating and flowing smoothly and help to alleviate pain symptoms.

Here are four ways to stay fit and make exercise a safe part of your sickle cell crisis prevention.

**Remember to rest when you feel tired and always consult with your doctor first before starting or changing your fitness regimen.**

[BLACKDOCTOR.ORG](http://BLACKDOCTOR.ORG)





## DRINK LOTS OF WATER

Dehydration - a loss of fluid - can slow down the blood flow in the body, increasing the chance of having a pain episode. Stay well hydrated at all times, but particularly before, during and after a workout.

## YOGA POSE FOR THE PAIN

Stress can be a sickle cell pain trigger and the practice of yoga is proven to relieve stress in the mind, body and spirit. Learning to breathe properly is a large part of the yoga practice and the increased oxygen intake helps the circulation of blood. Bikram "hot yoga" may be all the craze, but it is NOT for you if you have sickle cell. Stick to a gentle flow like yin yoga, restorative yoga or hatha.

## TAKE A WALK

The CDC recommends 150 minutes of moderate activity per week, and research has shown there are several health benefits to walking at least 30 minutes a day. If keeping a slow, steady pace, oxygen consumption will also stay steady. Walking on flat terrain is less taxing on the body, so treadmill walking is a good option since you can control the incline (unlike outdoor walking).

## SWIMMING & WATER AEROBICS (HEATED)

Swimming/Water aerobics (heated): Cold air and cold water is shocking to the body and can bring on a SCD pain crisis. However, even with sickle cell you can enjoy the benefits of low-impact water aerobics and swimming in an indoor heated pool. Before entering the pool, take a warm shower to avoid a sudden change in body temperature that can bring on a pain episode. Wrap yourself in a towel immediately after getting out of the pool.



# FINDING SUPPORT FOR SICKLE CELL

If you or someone in your family has sickle cell anemia, you may want help with the stresses of this lifelong disease. Sickle cell centers and clinics can provide information and counseling.

Ask your doctor or the staff at a sickle cell center if there are support groups for families in your area. Talking with others who are facing the same challenges you are can be helpful.

It's especially important to find ways to control — and cope with — pain. Different techniques work for different people, but it might be worth trying heating pads, hot baths, massages or physical therapy.

Prayer, family and friends also can be sources of support.



If you have a child with sickle cell anemia, learn as much as you can about the disease and make sure your child gets the best health care possible.

A child with sickle cell disease has special needs and requires regular medical care. Your doctor can explain how often to bring your child for medical care and what you can do if he or she becomes ill.



Living with sickle cell can be very challenging, but by working with your doctor to develop an effective treatment plan, as well as making certain key lifestyle adjustments, you can get a better handle on the disease — and the rest of your life.



# Create a SPECIALISTS YOU SHOULD KNOW Health Team

## Specialists You Should Know:

Creating a support system is important. Here are some specialists to include on your all-star health team when living with Sickle Cell Disease.

### 01 Ophthalmologist

Monitor and prevent eye complications with regular visits. Ophthalmologists should be seen each year or every 6 months--more often if doctor deems necessary.

### 02 Pulmonologist

It's important to undergo regular pulmonary function tests.

### 03 Cardiologist

See a cardiologist as needed when symptoms occur. If there are no symptoms it's best to at least visit for a baseline stress test

### 01 Orthopedist

If you have AVN or deep bone pain visit an orthopedist and have everything checked out.

### 02 Circulation

Avoid tight fitting or overly snug clothing. It hinders your circulation and can make you overheat.

### 03 Gastroenterologist

GI doctors are commonly seen for gall stones, a common complication for SCD patients



# Hydration Station

How do you know how much water you need? The old eight, eight-ounce glasses per day does not always apply. You may need more according to your level of activity.

Monitor your hydration by the paleness of your urine. The more hydrated you are, the more pale your urine will be-- you'll want your urine to be very pale.

Your eyes can also tell how hydrated you are. Do they appear a bit sunken in? You may need more water.

Add fruit juices, coconut water to replenish electrolytes. Excess sugar or alcohol consumption can dry out your cells or make them sticky.



# TIPS FOR THE EMERGENCY ROOM

1

**Emergency Kit:** Pack a tote or gym bag with things you may need on an ER visit such as a blanket, medication list, a pill bottle with a three day supply of your daily meds, heat pack, handi-wipes or sanitizer, toiletries, healthy snacks, and bottle of water or juice.

2

Even if you are able to, it's best not to consume snacks while waiting in the ER. Your condition or treatment may depend on it.

3

Pack a charger or battery pack for your phone and a book to read or puzzle to keep busy, or distracted from worry.

4

Waits in the ER can be longer than they should and you can become dehydrated in the process, make sure you have a supply of water. If you are running a fever, keep Tylenol with you. Ask a nurse first if its okay to drink or take medicine. Tell staff what medicine you recently took.

5

Wear a mask in the ER during flu season even if you don't have a cold. It will protect you from others who do.

LIVING WITH:

# SICKLE CELL DISEASE

*A GUIDE TO DAILY LIVING*

