

# LIVING WELL WITH SICKLE CELL!

<sup>TM</sup>

**UNDERSTANDING &  
KEEPING ON TOP OF  
YOUR DISEASE!**



# DON'T PANIC!

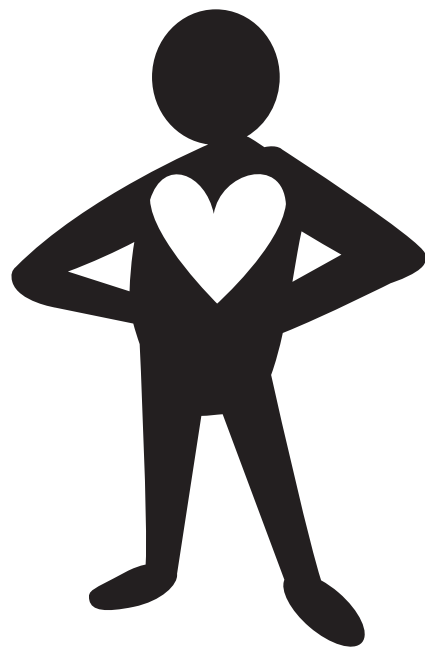
Most likely you're reading this book because you or someone close to you has been diagnosed with sickle cell disease. This can be a time in which you feel worried or scared. It's understandable. But a better way to feel better is to try to understand more about the disease and what you or they can do to "Live Well with Sickle Cell."

In this book you'll find out about what sickle cell is and how a person with the disease can do things to feel the best he or she can. Learning more about sickle cell can ease your mind and help you look at areas that are the most important to you so you can find additional information.

So don't panic. It may be a new chapter in your life, but it's far from the end.

## **DISCLAIMER.**

This book provides general information about sickle cell disease and related issues. The information does not constitute medical advice and is not intended to be used for the diagnosis or treatment of a health problem or as a substitute for consulting a licensed health professional. Consult with a qualified physician or health care practitioner to discuss specific individual health needs and to professionally address personal medical concerns.



# LIVING WELL WITH SICKLE CELL

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# THE BASICS OF SICKLE CELL DISEASE

Here are some common questions about sickle cell with plain answers.



## Q: WHAT IS SICKLE CELL?

A: Sickle cell disease is an inherited blood disease. You are born with it and it lasts a lifetime.

## Q: WHAT ARE THE SIGNS AND SYMPTOMS OF SICKLE CELL?

A: They're different. Some people have mild symptoms. Others have very severe symptoms and often are hospitalized for treatment.

## THE MOST COMMON SIGNS AND SYMPTOMS OF SICKLE CELL DISEASE ARE:

Anemia

Pain when sickle-shaped red blood cells block blood flow and oxygen

Other more specific symptoms

Tiredness, lack of energy

Paleness

Yellowing of the skin and eyes (jaundice)

Shortness of breath

Prone to infections

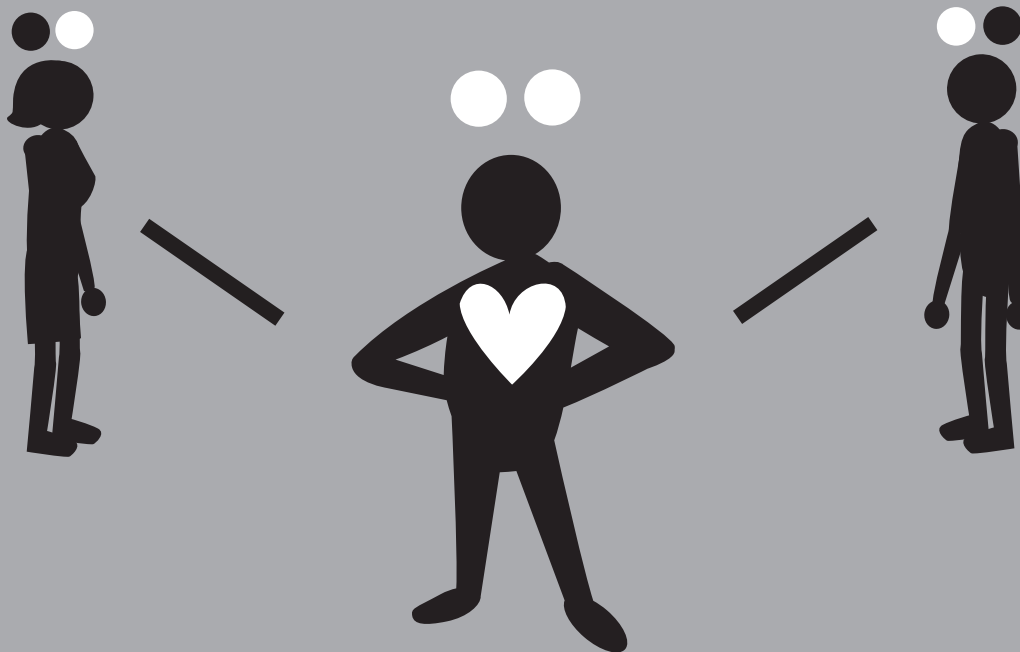
Liver and kidney damage

Difficulty breathing

Skin ulcers and sores on the lower legs

## Q: WHAT CAUSES SICKLE CELL DISEASE?

A: People with sickle cell disease inherit two genes, one from each parent, that are different from normal genes (variant). These variant genes are called sickle cell genes.



## Q: WHO GETS SICKLE CELL?

A: Anyone can have sickle cell disease:

Children  
Adults  
Elderly

## Q: HOW IS SICKLE CELL DIAGNOSED?

A: Screening tests are done on newborn infants. These tests show if the newborn infant has sickle cell disease or carries the cell trait.

## Q: HOW IS SICKLE CELL TREATED?

A: By relieving pain, preventing infections and controlling any complications. The treatments include:

Medications  
Blood transfusions  
Specific treatment for complications

# FAST FACTS ABOUT SICKLE CELL DISEASE

## QUICK HITS TO GET YOU UP TO SPEED

Sickle cell is a blood disease that you are born with that lasts a lifetime.

It is a disorder that affects the red blood cells.

The disease is inherited, meaning that your mother and/or father carry the trait or have the disease.

Sickle cell is not contagious. You can't catch it from sneezing, coughing or kissing. You have to be born with the gene for it, passed from your parents.

Normal red blood cells can live for 120 days, but sickle-shaped cells live only 10 to 20 days.



NORMAL RED  
BLOOD CELL



SICKLE RED  
BLOOD CELL

In the US, sickle cell most commonly affects African-Americans. About one out of every 400 African-American babies born in the US has sickle cell disease. But it can also affect all other races and ethnicities.

There are effective medicines and treatment plans for dealing with sickle cell.

## MISSION: DEFINITION

**Anemia (uh-nee-me-uh):** a deficiency of the hemoglobin, often accompanied by a reduced number of red blood cells and causing paleness, weakness and breathlessness.

# LIVING WITH SICKLE CELL

## WHAT TO DO

First things first. If you have sickle cell disease, it's important to take good care of yourself.

### THINGS YOU NEED TO DO TO TAKE CARE OF YOUR HEALTH:

- Maintain a healthy diet.
- Drink water every day.
- Get enough sleep and rest.
- See your doctor regularly for checkups and treatment.
- Get a flu shot.
- Educate yourself about your disease.
- Avoid stress and extreme heat or cold.

## THE TIP-OFF SICKLE CELL BY THE NUMBERS

1 in 600  
African-Americans have  
sickle cell disease

1 in 1,000-1,400  
Hispanic-Americans have  
sickle cell disease

1 in 12  
African-Americans have  
the sickle cell trait

Sickle cell disease affects  
up to 100,000 Americans

Over 3.5 million African  
Americans are carriers of  
the sickle cell trait

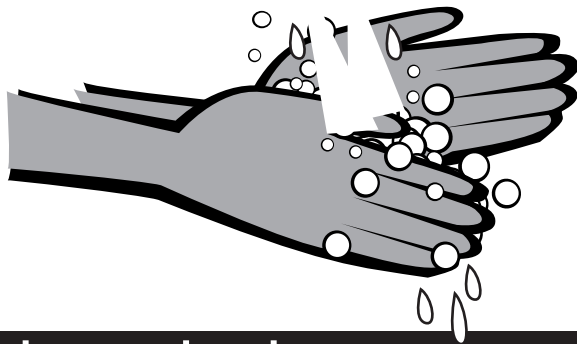
**CHECK IT OUT!**   
[sicklecellwarriors.com](http://sicklecellwarriors.com)

# THE INFECTION CONNECTION

## FIVE TIPS TO PREVENTING INFECTION

Everyone gets sick with colds, flu and viruses from time to time. For people with sickle cell, these illnesses can quickly become dangerous. But you can take some simple measures to protect yourself. Here are five steps to take to help keep you healthy and infection-free.

### TIP #1: WASH YOUR HANDS



It's so easy. It takes only a few seconds. And it's one of the best ways to prevent getting infections. People with sickle cell, their families and their caregivers should wash their hands regularly. Use soap and water. Also, keep gel hand sanitizer close by and use it often.

#### Wash your hands...

Before making food

Before eating

After using the bathroom

After blowing your nose, coughing or sneezing

After touching people or things that can carry germs, such as:

Diapers or children who have used the toilet  
Uncooked food (raw meat, eggs, unwashed veggies)

Animals or animal waste

A sick person

#### BETCHA DIDN'T KNOW

Antibacterial soap is no better at killing germs than regular soap.

Using antibacterial soap may even create bacteria that are resistant to the product's antimicrobial agents, making it harder to kill these germs in the future.

CHECK IT OUT! 

[cdc.gov/handwashing/why-handwashing.html](http://cdc.gov/handwashing/why-handwashing.html)



## TIP #2: KEEP YOUR FOOD SAFE



### Beware of Bacteria

Keeping your food clean, fresh and safe from germs is seriously important. The salmonella bacteria that can affect some foods can be dangerous and harm children with sickle cell.

### How to Stay Safe When Cooking and Eating:

Wash your hands, cutting boards, counters and all utensils after they touch uncooked food.

Before you eat them, wash fruits and vegetables well.

Cook meat until it's well done. The juices should run clear with no pink inside the meat.

Never eat raw or undercooked eggs. Be extra careful of uncooked eggs that may be in homemade hollandaise sauce and salad dressing, homemade ice cream, homemade mayonnaise, cookie dough and frosting.

Don't eat raw or unpasteurized milk or other dairy products, like cheese.

**GOOD ADVICE:** Always look for the word **PASTEURIZED** on the label of dairy products like milk and cheese.

### CHECK IT OUT

If you want to see if an egg is fresh, take it for a swim. Put the eggs in a bowl of clean water. Fresh eggs will sink down and lie on their sides at the bottom of the bowl. Semi-fresh eggs will stand on end and bob in the water. Old eggs will float. Throw those away immediately. This is because of the air inside the shell of the egg. Older eggs have larger air pockets and cause them to float.



FRESH



SEMI-FRESH



OLD

CHECK IT OUT!



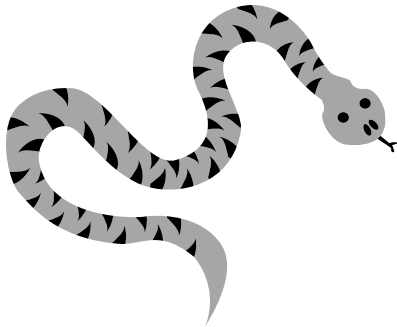
[foodsafety.gov/](http://foodsafety.gov/)

# TIP #3: AVOID REPTILES

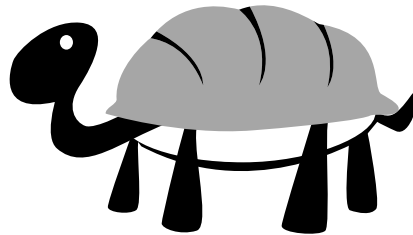
## Scale Models

Most people get a little freaked out by the sight of a snake. They're creepy and scary and slither around and some are venomous and deadly. But children with sickle cell disease have an added danger when it comes to snakes and other reptiles. Some reptiles carry the salmonella bacteria, which is especially harmful.

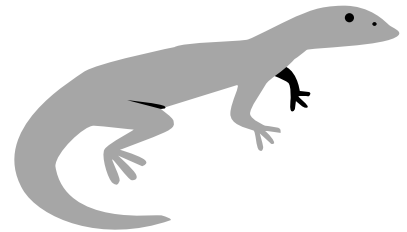
To avoid salmonella contact, children with sickle cell disease should always stay away from:



Snakes



Turtles & Tortoises



Lizards

## MISSION: DEFINITION

**Salmonella:** Any of several rod-shaped, anaerobic bacteria that may enter the digestive tract of humans and other mammals through contaminated food, causing abdominal pains and diarrhea.

Alligators and crocodiles are reptiles, too. But they should probably be avoided for a whole other reason!

CHECK IT OUT! 

[salmonella.org/info.html#reptiles](http://salmonella.org/info.html#reptiles)

# TIP #4: VACCINES

## A Good Defense

Vaccines prevent many serious childhood illnesses and infections. Children with sickle cell should receive all the regular childhood vaccines, plus a few extra, in order to reduce the risk of getting these ailments.

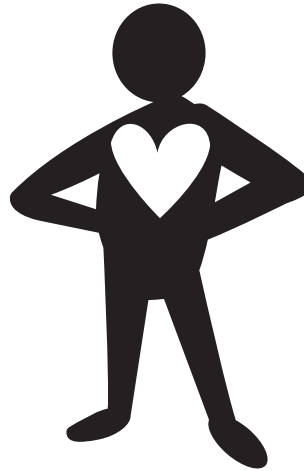
### EXTRA VACCINES:

A flu vaccine every year after six months of age.

A pneumococcal vaccine at 2 and 5 years old.

Meningococcal vaccine, for some children at risk.

The flu vaccine is recommended each year for all adults with sickle cell as well. Adults should also get the pneumococcal vaccine and any others recommended by their doctors.



## THE TIP-OFF

Don't Mean to Scare You But...

People with sickle cell disease are more likely to contract pneumococcal infection, which may be fatal. Medical researchers are currently hard at work to develop sophisticated vaccinations that will be effective for preventing pneumococcal infection for all ages.

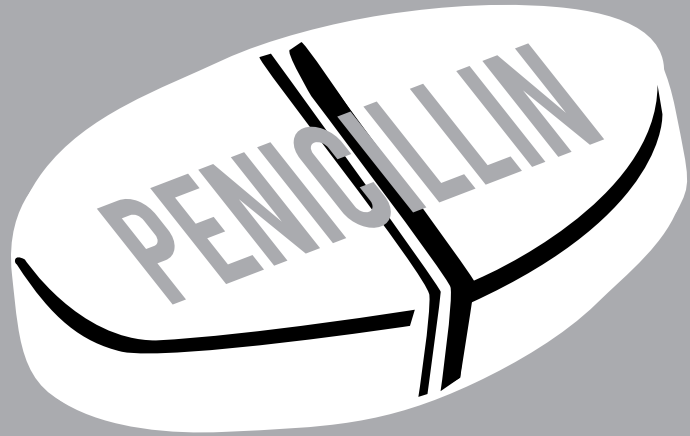
CHECK IT OUT! 

[vaccineinformation.org/pneumchild/qandavax.asp](http://vaccineinformation.org/pneumchild/qandavax.asp)

# TIP #5: PENICILLIN

## MISSION: DEFINITION

**Penicillin:** The most famous and popular of the antibiotics, it is derived from the fungal mold and named after the Latin *Penicillium notatum*. Penicillin is effective because it destroys bacteria's cell walls.



## Fighting for You

Penicillin is an effective tool in the fight against infection. If you're at risk for contracting infection-causing bacteria, you want this guy on your team. Young children are at extreme risk, so they're often prescribed penicillin every day until at least the age of five.

## BETCHA DIDN'T KNOW

Penicillin got its name from the Latin "penicillium," meaning "a painter's brush." Early scientists thought the fronds of the fungus looked like a painter's brush.



**CHECK IT OUT!**



[medterms.com/script/main/art.](http://medterms.com/script/main/art.asp?articlekey=15277)

[asp?articlekey=15277](http://medterms.com/script/main/art.asp?articlekey=15277)

# YOU'RE NOT ALONE

## FAMOUS PEOPLE WITH SICKLE CELL

Many people have suffered from sickle cell disease and gone on to accomplish great things and fulfill their dreams. You can too.

Tionne “T-Boz” Watkins: Singer from the group TLC

Larenz Tate: Actor

Miles Davis: Jazz Musician

Tiki Barber: New York Giants football player

Paul Williams: Singer with the Temptations

Georgeanna Tillman: Singer with the Marvelettes

Prodigy: Rapper, Mobb Deep

CHECK IT OUT! 

[childrensnational.org/Sickle-Cell/ask-an-expert.aspx#Q2](http://childrensnational.org/Sickle-Cell/ask-an-expert.aspx#Q2)

### CHECK THIS OUT

Sickle cell disease is far from new. The people of Africa have known of the condition for hundreds of years. In West Africa, various native ethnic groups called it by different names:

Ga tribe: “chwechweechwe”

Faute tribe: “nwiiwii”

Ewe tribe: “nuidudui”

Twi tribe: “ahotutuo”

# THE SICKLE CELL SIX

## HEALTHY TIPS FOR LIVING WELL WITH THE DISEASE

Do you have sickle cell disease? Do you have a family member or friend with the condition and want to help? Like the people mentioned on the previous page, there is no reason why you can't live a full, active life with sickle cell. Here are six helpful tips to help you or someone you know with sickle cell stay as healthy as possible.

### 1 Find good medical care.

Because sickle cell is a complicated disease, finding good quality doctors and nurses is really important. Your doctors and nurses should know everything there is to know about your condition and should be able to help prevent serious problems. A hematologist working with a group of highly trained specialists is what you want on your team.

### MISSION: DEFINITION

**Hematologist:** A doctor specializing in blood diseases.

### 2 Get regular checkups.

To prevent serious health problems, schedule regular appointments with a primary care doctor. These routine visits can find potential problems and address them before they become more serious and complicated.

|  |                                       |
|--|---------------------------------------|
| Babies from birth to 1 year              | See a doctor every 2 to 3 months      |
| Children from 1 to 2 years               | See a doctor at least every 3 months  |
| Children at least 2 years old and adults | See a doctor at least once every year |

### **3 Prevent infections.**

Getting sick is a total drag. Taking some simple steps to help you keep from getting infections should be your highest priority. For a child with sickle cell, common illnesses like a cold and the flu can quickly become dangerous. Be smart and be prepared.

### **4 Learn healthy habits.**

Adopting a healthy lifestyle is crucial for people with sickle cell. Here are some simple daily habits to remember:

DRINK 8 TO 10 GLASSES OF WATER EVERY DAY

EAT HEALTHY FOOD

DON'T LET YOURSELF GET TOO HOT, TOO COLD OR TOO TIRED

PARTICIPATE IN PHYSICAL ACTIVITY BUT DON'T OVERDO IT

REST WHEN TIRED

## 5 Look for clinical studies.

In an effort to understand and better treat the disease, researchers conduct new clinical studies all the time. The hope is that these studies will eventually find a cure for sickle cell disease, or at least more effective treatments. People who participate in these studies might have access to new medicines and treatment options.

## 6 Get support.

You can't do it alone and you don't have to. Patient support groups and community-based organizations that can give you information, assistance and support are out there. Find them. They're there to help.

These six tips for living well can help you lead an active life and stay healthy. If you're a friend or family member to someone with sickle cell, learn these tips and understand what he or she is dealing with. You too can make a big difference and be a huge help. Be understanding. Be helpful. Be a friend.

**CHECK IT OUT!**   
[cdc.gov/ncbddd/sicklecell/healthy-living-living-well.html](https://cdc.gov/ncbddd/sicklecell/healthy-living-living-well.html)



# **CODE RED**

## **EMERGENCY SITUATIONS**

### **When to See a Doctor**

Because sickle cell is a very serious condition, emergency situations can arise at any time. It's important that every person or family with a young child with the disease have a plan in place for getting care immediately, day or night, should there be a problem. If the care facility is new to you and doesn't have your records on file, make sure you bring a copy.

### **GO TO AN EMERGENCY ROOM OR URGENT CARE FACILITY IMMEDIATELY FOR:**

Fever above 101 degrees

Difficulty breathing

Chest pain

Abdominal (belly) swelling

Severe headache

Sudden weakness or loss of feeling and movement

Seizure

Painful erection of the penis that lasts more than four hours



### **CALL A DOCTOR RIGHT AWAY FOR:**

Pain anywhere in the body that won't go away with home treatment

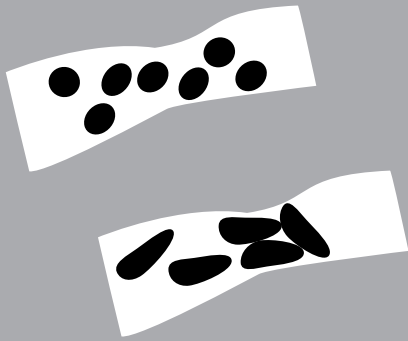
Any sudden problem with vision or eyesight

# WHAT'S GOING ON HERE?

## THE SICKLE CELL CRISIS

### Go with the Flow

Normal, healthy red blood cells are round and smooth. Their shape lets them easily pass through tiny blood vessels. But people with sickle cell disease have abnormal, elongated, sickle-shaped red blood cells. These cells are hard and sticky. Their shape makes it difficult to flow through the vessels, often clogging them and preventing blood flow. This clog is called an “occlusion.” An occlusion in the vessel can create a very painful sickle cell crisis.



### The Crisis Breakdown

About **30%** of sickle cell sufferers

Rarely or never have the pain caused by a crisis.

About **50%** of sickle cell sufferers

Experience only a few severe crises throughout their lives.

About **20%** of sickle cell sufferers

Have frequent and severe sickle cell crises.

For those who do experience sickle cell crises, the following are factors that may cause a crisis to begin:

Being in cold weather

Feeling exhausted or fatigued

Exercising too hard

Being dehydrated from not drinking enough water and other fluids

Not having enough oxygen in the blood

Sometimes a crisis gets so painful that a trip to a hospital's emergency room is necessary in order to receive immediate treatment with pain medicine.

**CHECK IT OUT!**   
[emedicinehealth.com/  
sickle\\_cell\\_crisis/article\\_em.htm](http://emedicinehealth.com/sickle_cell_crisis/article_em.htm)

# WHY ME?

## THE SICKLE CELL TRAIT AND YOUR GENES

### MISSION: DEFINITION

**Hemoglobin:** The oxygen-carrying pigment of red blood cells that gives them their red color and serves to send oxygen to the tissues.

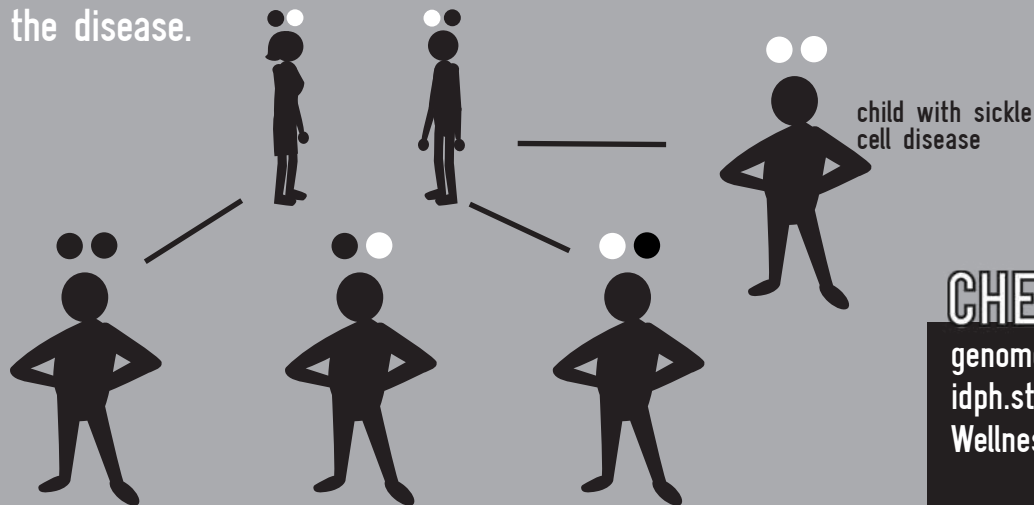
Sickle cell is an inherited disease. It's passed from parents to their children (heredity). Every baby born gets a gene from his father and one from his mother. These genes decide the make-up of the hemoglobin. If you're in a family with a history of sickle cell disease, there are three possibilities:

1. You have two genes for healthy hemoglobin. You will have no signs of sickle cell.
2. You have one gene for healthy hemoglobin and one gene for sickle hemoglobin. This is known as having the "sickle cell trait." If you have the sickle cell trait, you are not sick and probably don't even know you have the gene, unless you've been tested.
3. You have two genes for sickle hemoglobin, and have sickle cell disease.

### Straight to the Trait

Sickle cell trait is a condition in which there is one gene that forms sickle hemoglobin and one that forms normal hemoglobin. Sickle cell trait occurs in one out of every 12 African-Americans. Usually, people with sickle cell trait do not have any medical problems and they can lead normal lives. They do not develop sickle cell disease.

Before having children, people with the disease or with a family history of sickle cell may want to speak with their doctor to find out how likely a child is to be born with the disease.



**CHECK IT OUT!**

[genome.gov/10001219](http://genome.gov/10001219)  
[idph.state.il.us/HealthWellness/sicklecell.htm](http://idph.state.il.us/HealthWellness/sicklecell.htm)

# WHAT TO DO ABOUT IT

## TREATMENT AND MEDICINE



### The Bad News

The only known cure for sickle cell disease currently is a bone marrow transplant, which requires highly specialized care from a hematologist who performs these transplants.



### The Good News

There are treatments and precautions to help treat the disease, prevent painful sickle cell crises and relieve the pain brought on by the condition. As mentioned before, you can avoid some of these crises by staying out of the cold, drinking plenty of water, getting rest and not exercising too hard.

### Some General Treatments

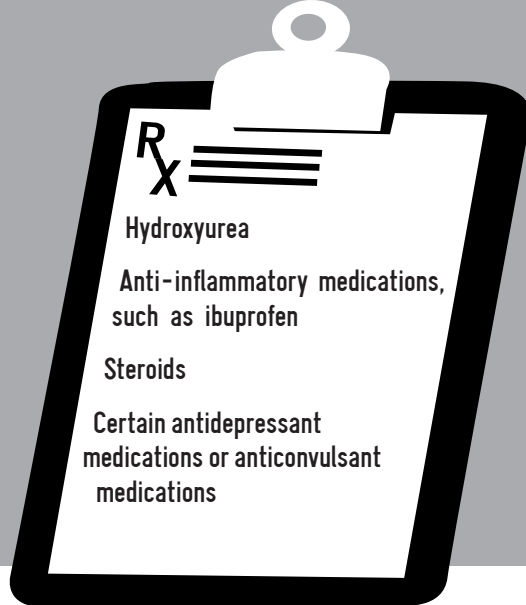
Your doctor may prescribe a medication such as erythropoietin to help with anemia. This is given as a shot and improves the blood count. Blood transfusions are sometimes used to improve anemia, as well.

While addiction is a concern with opioid medications, they can be extremely useful during painful sickle cell crises. Patients who have multiple crises can sometimes benefit from taking opioid medications daily, along with additional pain medication during crises. The daily opioids can help reduce the number of crises and lessen the pain.

Hydroxyurea is also sometimes prescribed for certain patients. This is a medication that might reduce the chance that the red blood cells will change into the sickle shape.

Doctors might also recommend that sickle cell patients take extra vitamins, especially folic acid.

Other medications that may be prescribed by a doctor for relief from sickle cell pain are:



## THE TIP-OFF

Many people find that they have problems taking the medications prescribed for them. Use the tips below if you have had some problems in the past.

Don't stop taking or change your medications without talking with your doctor first.

Tell your doctor right away if you're having problems with your medications.

Tell your doctor if you notice any new symptoms or side effects. Use a pill box to remember to take medications at the right time everyday.

Ask your doctor what to do if you forget to take a dose of medicine at the right time.

Identify cues in your daily life to help remind you to take your medicine.

## CHECK IT OUT!



[mayoclinic.org/diseases-conditions/sickle-cell-anemia/diagnosis-treatment/drc-20355882](http://mayoclinic.org/diseases-conditions/sickle-cell-anemia/diagnosis-treatment/drc-20355882)

Plan ahead for times when your routine may be different, like on vacations, holidays or weekends.

## Nondrug Treatments

Sometimes it's helpful for people with sickle cell to see a physical therapist. These specialists use gentle exercises, massage and heat and cold treatments to help manage the pain associated with sickle cell.

Seeing a psychologist, psychiatrist or social worker can also benefit those afflicted with the disease. Getting help for coping with the illness or learning techniques for controlling pain can be discovered through these specialists. Feeling frustrated? Confused? Alone? Talk to someone who can help. Don't be afraid to reach out.

As mentioned earlier, in severe cases, it's also a possibility for hematologists (blood and bone marrow specialists) to perform a bone marrow transplant. This usually requires healthy bone marrow from a brother or sister of the patient. This operation requires highly specialized care, but recent advances have made this an effective treatment, with the proper care and medications.

# NOT THE END

## LIVING WELL WITH SICKLE CELL

### Summing Up

Because of new advancements and research in the fight against sickle cell disease, many patients are living longer, fuller lives today. Take the precautions discussed in this book and maintain a regular relationship with your doctors and nurses to ensure a rewarding life where you keep your pain in check.

### Dare to Care

Because sickle cell disease is a serious condition, the family members of people with it are also affected by the illness. It may be stressful to deal with a child or loved one who has special health needs and requires frequent trips to the hospital. Parents and other family members may feel frustrated with the health care system and the schools for their lack of understanding about the illness. Be patient and know these family members care and are trying to help. If you're the parent or family member, try to understand what your loved one is feeling and experiencing in fighting the disease. The lives of everyone involved will be richer for it.



CHECK IT OUT!



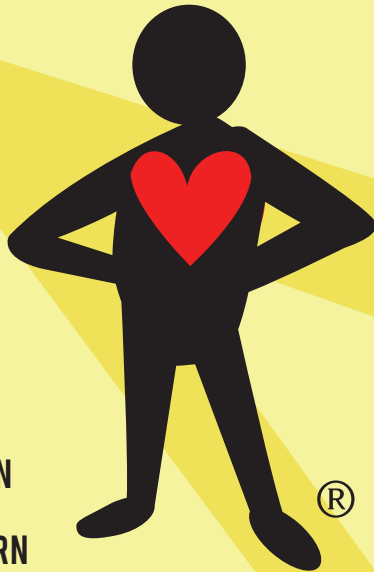
[ehow.com/how\\_4718369\\_live-sickle-cell-anemia.html](http://ehow.com/how_4718369_live-sickle-cell-anemia.html)

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# LIVING WELL WITH SICKLE CELL!

™

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DESIGN BY:  
SAM WASHBURN

**ACKNOWLEDGEMENTS FOR LIVING WELL WITH SICKLE CELL**  
Sickle cell can be devastating for children and families coping with this disease. Across the country, high rates of hospitalizations and emergency room visits are common for those suffering with sickle cell. Children's National Medical Center works closely with sickle cell patients, schools, families, and health care providers to educate about the disease, and provide access to health care services to prevent unnecessary hospitalizations. Families need caring and knowledgeable health care providers who are committed to outreach and educating patients to equip them with the tools they need to combat sickle cell. The Child Health Advocacy Institute at Children's National believes the "Living Well With Sickle Cell!" is a wonderful tool for health care providers and families affected by Sickle Cell Disease. We appreciate Centene Corporation's leadership on this important health issue.

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Children's National Medical Center  
Washington, DC



MADE IN  
THE U.S.A.



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