

A GUIDE TO LIVING WITH
SICKLE CELL DISEASE

DEFINE YOURSELF, *Define your life*



Greetings Warrior,

This booklet was inspired, designed and created by your brothers and sisters in the sickle cell community to encourage, uplift and inform you. We want you to know you are not alone. **Sickle cell is part of our lives, but it does not define us and it cannot break us.** Join us as we redefine what it means to live with sickle cell.

If you are interested in learning more about this project, please visit: www.sc101.org/litproject.

Cassandra Trinnell

Cassandra Trinnell
Project Manager,
Sickle Cell Community Consortium
President, Sickle Cell 101

Lakiea Bailey

Lakiea Bailey, Ph.D.
Executive Director
Sickle Cell Community Consortium

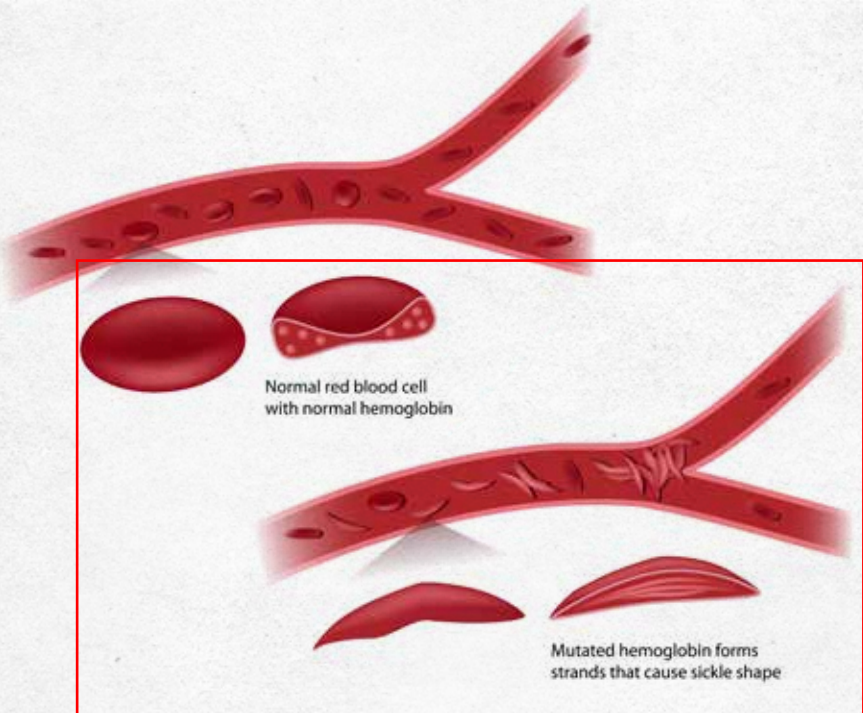


DEFINE YOURSELF, *Define your life*

No matter how old you are or how severe your sickle cell disease is, you can start where you are and learn more about sickle cell disease. The more you know, the more options you have and more options exist now than ever before. Advances are being made, and there is hope.

Defining **SICKLE CELL DISEASE**

Sickle cell disease is a life-long, genetic blood disorder that affects how oxygen is carried in the body. Red blood cells are round and flexible and contain a protein called hemoglobin that delivers oxygen easily throughout the body. Sickle cell disease causes red blood cells to become hard, sticky and take on a sickle, or crescent, shape. Sickle cells get stuck in blood vessels and block blood flow to organs like kidneys, liver, lungs and heart. When your organs don't get the oxygen they need, you may feel intense pain, which is known as a vaso-occlusive pain crisis, pain crisis or pain episode.



Types of SICKLE CELL DISEASE

HbSS

HbSC

HbS_{beta}
thalassemia

- Hb = hemoglobin
- S = sickle gene
- C = abnormal hemoglobin

All of these types are known collectively as sickle cell disease.

Sickle cell disease can affect people from all ethnic backgrounds, but in the United States it primarily affects African-Americans. Sickle cell disease is an inherited genetic disease which means you have two sickle cell genes, one from each parent. For example, if you have sickle cell anemia, you have inherited one sickle cell gene ("S") from your mother and another sickle cell gene ("S") from your father. If you have HbSC, you have inherited one sickle cell gene ("S") from one parent and an abnormal hemoglobin gene ("C") from the other parent. The combinations of genes you inherit from your parents determine the type of sickle cell disease you have. If you don't know what type of sickle cell disease you have, ask your doctor so you know what treatment options are available.

There also are a few rare types of sickle cell disease: HbSD, HbSE, and HbSO. The severity of these rarer sickle cell disease types varies.

Sickle Cell Trait

Sickle Cell Trait is found in people who inherited one sickle cell gene ("S") from one parent and one normal gene ("A") from the other parent. People with sickle cell trait may not have any symptoms of sickle cell disease, but in some cases may experience complications such as pain crises. It is important to be tested for the trait because sickle cell trait can be passed on to your children. If both you and your partner have sickle cell trait you have a 25% chance of having a child with sickle cell disease with each pregnancy. Also, if you have sickle cell trait, ask your doctor about renal medullary carcinoma, a **rare** aggressive cancer more commonly found in people with the sickle cell trait.

HbAS

- Hb = hemoglobin
- A = normal gene
- S = sickle gene

SICKLE CELL DISEASE

and Your Body

Sickle cell disease affects everyone differently and the complications can range from mild to severe.

PAIN: The most common complication, pain, can be acute or chronic.

Acute Pain: When the sickle cells get stuck in blood vessels anywhere in the body it can lead to sudden, severe acute pain. Acute pain also is known as a vaso-occlusive crisis.

Chronic Pain: As people living with the disease grow older, many suffer from ongoing, chronic pain. The cause of chronic pain is not well understood, but it may be related to organ damage.

ORGAN, TISSUE, BONE PAIN AND

DAMAGE: Pain or damage can occur in organs, tissues or bones when that part of the body is deprived of oxygen due to sickle red blood cells blocking the blood flow.

ANEMIA: This occurs when the number of red blood cells is low. Severe anemia may cause pale skin, difficulty breathing, tiredness or dizziness. In children, anemia may be due to blood trapped in the spleen (which is an emergency) or by a viral infection. Seek medical attention if you have these symptoms.

INFECTIONS: A common cold, flu or bacterial infection can quickly

My acute pain usually happens when I wake up in the middle of the night in crisis. As I've gotten older my bones have started hurting all the time, day and night. This is chronic pain. -L.B.

become more serious with sickle cell disease. Sickled cells can damage the spleen, which produces white blood cells that are critical in protecting your body against certain germs.

ACUTE CHEST SYNDROME: This severe complication of sickle cell disease causes lung damage leading to chest pain, fever, intense coughing and sometimes lung infection. Acute chest syndrome may be associated with a pain episode, especially in adults, and may quickly become very serious.



PREVENTION

Knowing your body and what may trigger a complication will help you manage your sickle cell disease and give you the power to define your life.

RELAX: Stress can lead to complications for those living with sickle cell disease. Make time for activities that help you relax and reduce stress like meditation, reading, walking or spending time with loved ones.

I recommend that you do your best to live toxic-free. Read the labels of the foods you eat, know what the ingredients mean. Eat foods that support longevity. -T.J.

SEE YOUR HEALTH CARE TEAM REGULARLY:

Be proactive when symptoms occur, keep regular appointments so your health care team knows what is normal for you and can provide preventative care.

Making healthy choices is more important when you are living with sickle cell disease and may even prevent pain crises and other complications. Eating right, exercising, drinking plenty of water and getting enough

One of the scariest parts of having a baby diagnosed with sickle cell is having to figure out when your child is sick or in pain. Never be afraid to seek medical attention if you are slightly concerned. Keep your child hydrated and don't worry if they start wetting the bed. Bed wetting is common in this disease. -D.B.

sleep are all ways to improve your overall health. Washing hands frequently and getting immunized will protect you from infections. Minimize unhealthy choices, such as cigarettes and alcohol, and focus on making wise decisions.

My major triggers are cold weather and stress. I've learned that I stay healthier for longer periods of time if I avoid them both. - L.J.

Defining **YOUR TRIGGERS**

Triggers, such as extreme heat or cold, stress, dehydration and overexertion, may cause a pain crisis or other complications. Know your triggers so you can work with your healthcare team to anticipate and manage the disease.



Treatment **OPTIONS**

Knowing your triggers can prevent some pain crises, but currently not everything is avoidable. If a crisis or complication occurs, it can be stressful and hard to manage. Talk with your healthcare team to see what treatment options below might work best for you.

DISEASE MANAGEMENT AND TREATMENT OPTIONS

Blood transfusions/exchange transfusions: This delivers healthy red blood cells to the body to help prevent complications.

Hydroxyurea: This oral medicine may reduce the number of pain crises for some.

Pain medications: Nonsteroidal anti-inflammatory drugs (NSAIDs), opioids, antidepressants and anticonvulsants can help manage pain.

ONE-TIME CURATIVE OPTION

Stem cell transplant: During this one-time procedure, stem cells are taken from the bone marrow of a healthy donor who is a genetic match, usually a sibling, and transplanted into the person with sickle cell disease to create a supply of new stem cells that are able to make normal red blood cells.

INDIVIDUAL *Home Treatment Plan*

Talk to your doctor about creating an individualized home treatment plan, a written, personal plan to help you prevent and control pain and complications at home. Your plan might include drinking water, taking pain medicines, using a heating pad or meditating. Update your plan regularly, refer to it when necessary and take it with you to the emergency room.

Clinical Research Studies

YOU MAY HELP DETERMINE FUTURE TREATMENTS

Clinical research studies are conducted by doctors to see if a possible new treatment improves health. Participating in a clinical study may be an option to consider if you want to contribute to the research process, if the standard treatment options no longer work for you or if you want a different option. Talk with your doctor to see what clinical study options may be available for you. By participating in a study, you may help determine future treatments for sickle cell disease.

EXPERIMENTAL TREATMENTS

Researchers are conducting many clinical research studies to better understand and develop possible treatments for sickle cell disease, pain management, and iron overload to treat symptoms and the disease. Additional curative options also are being studied:

Fetal hemoglobin production: Researchers are studying medicines that may reactivate the production of fetal hemoglobin, which prevents red blood cells from sickling. The hope is that reactivating the fetal hemoglobin will reverse the disease.

Gene therapy: This involves harvesting the patient's own stem cells that contain the genetic defect that causes the disease, modifying the cells in a lab to include corrected genes, and returning to the body so the modified cells may create a supply of healthy red blood cells.

ALTERNATIVE TREATMENT OPTIONS

Some people find pain relief from massage therapy, acupuncture, non-medicinal pain creams and oils, and meditation.

If there are alternative treatment options that work for you, please share them with us at www.facebook.com/group/SCDnatural



Ways to **IMPROVE YOUR LIFE WITH SICKLE CELL DISEASE**

Living with sickle cell disease impacts your body and your entire life. Managing your life with the disease goes beyond the doctor's office.

ADVOCATE FOR YOURSELF

Speak Up. You are the only person who can tell the doctor how you feel. You are the best expert on you. Be bold, speak up and be descriptive – clearly explain how you feel. This can help your treatment team provide you with the best care.

Keep Track. You may want to keep a journal and write in it each day to track how often and when you don't feel well.

Your educational goals can be as big as your dreams. We do not live on the timetable of others. What has helped me is realizing this, asking for help when I need it and making use of the disability office on campus. - M.B.

Seek Support. It is okay to seek support and to ask for help. Reach out to people you trust and allow your friends, family, or even co-workers and classmates, to support you on this journey.

Take your mental health as seriously as you take your overall health. In order to properly take care of your body, you have to also take care of your mind. Seeing a psychiatrist is part of my care plan and I love it! - S.W.

Take Care of Your Mental Health.

Your mental health is important. Let your doctor or a loved one know if you are not feeling like yourself. Resources exist and it is okay to seek help from a support group or therapist.

Communicate Clearly With Others.

You may decide not everyone needs to know about your disease, but it can help if you are open and honest about how you feel. Clearly communicating how you feel and what your limitations are may help others understand what you are experiencing.

Manage Your Healthcare.

Taking the time to get and stay organized will help you manage your treatment. Give yourself extra time to get places, organize your appointments and take time to make plans each night for the next day.

I've raised two children with sickle cell. This disease isn't the end of the world. Have patience and take it one day at a time. Show a lot of love and know that sickle cell is manageable. - A.E.

Learn more about the human body and its connection mentally, spiritually, physically and emotionally. These elements can interact with you in a negative or positive manner. Know the difference, sometimes the choices we unconsciously make may be hurting us. -T.J.

References:

Centers for Disease Control, <http://www.cdc.gov>
National Heart, Lung and Blood Institute, <http://www.nhlbi.nih.gov>

Things to **REMEMBER**

My diagnosis is: _____

My primary care physician is: _____

Contact info: _____

My hematologist is: _____

Contact info: _____

My blood type: _____

Allergies: _____

Daily medications: _____

My baseline hemoglobin count (baseline hemoglobin varies from person to person. It is important that I know my normal count when I am not in sickle cell crisis):



PATIENT-POWERED, PHYSICIAN REVIEWED

Sickle Cell Literature Project

PRESENTED BY:

SICKLE CELL 101

IN COLLABORATION WITH:



CONTENT REVIEWED BY:

The Sickle Cell Community Consortium Healthcare and Research Advisors

This project was funded by an educational grant from bluebird bio and with the support of Sickle Cell Community Consortium (SCCC) donors. To learn more about the SCCC and our patient-driven projects and priorities, visit www.sicklecellconsortium.org. The content was inspired, designed and created by the patients and families of the sickle cell community.

For more information about this project visit: www.sc101.org/litproject.