NATIONAL SICKLE CELL AWARENESS MONTH

SEPTEMBER

#SickleCellMatters

SCDAA

"Break The Sickle Cycle"®

September is National Sickle Cell Month

Toolkit

National Baptist Convention, USA, Incorporated
Dr. Jerry Young, President
Dr. Alvin Edwards, General Secretary
Dr. Alvin Love, Chair, Federal Faith-Based Initiative
Dr. Michael O. Minor, National Director, NBCUSA H.O.P.E. HHS Partnership

Sickle Cell Sabbath Sunday 2024
Toolkit Overview
Sunday, September 15, 2024

Edited By
Dr. Genice Nelson
NBCUSA H.O.P.E. National Sickle Sabbath Sunday Chair
info@hopenbc.com

Overview: What Is Sickle Sabbath Sunday?

Sickle Sabbath Sunday is a national faith day to raise awareness for sickle cell disease in the community and provide charitable giving within our congregations. As a national partner of the National Sickle Cell Disease Association of America (SCDAA), the National Baptist Convention, USA, Incorporated, encourages our member churches to conduct sickle cell awareness and education activities on Sickle Sabbath Sunday.

Facts about Sickle Cell Disease in the United States

Source: National Sickle Cell Disease Association of America

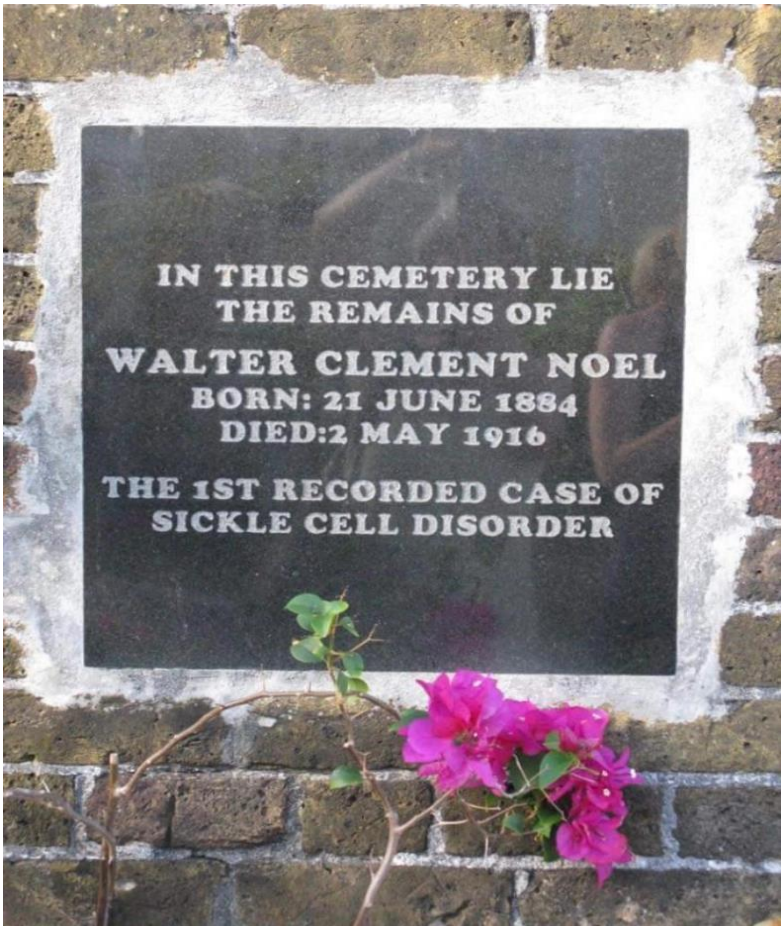
Sickle Cell Disease (SCD) is an inherited blood disorder which constitutes a global health problem. Approximately 100,000 Americans are living with the disease. One in every 400 African Americans and one in every 14,000 Hispanic Americans is born with SCD. Many more Americans perhaps as many as 2.5 million – including 1 in 10 African Americans are carriers for Sickle Cell Trait (SCT). Given that these carriers are generally unaffected by SCD complications, many are unaware that their children are at increased risk for the disease.

In November 2010, the world recognized the 100th year anniversary of the discovery of the very first genetic disease: Sickle Cell Disease.

Dr. James B. Herrick, an attending physician at Presbyterian Hospital and professor of medicine at Rush Medical College in Chicago, Illinois, published an article on the case of an anemic West Indian patient. That

patient Walter Clement Noel was a student from Granada studying Dentistry in Chicago. Although Dr. James Herrick is credited with this discovery-his intern Dr. Jeremy Irons is the individual that made this discovery by obtaining the peripheral blood smear that Dr. Herrick is credited with reporting as his clinical and laboratory findings of the patient’s "peculiar elongated and sickle-shaped” red blood corpuscles which represents the first description of sickle cell anemia in Western medical literature.

Walter Clement Noel completed his dental studies, returned to Granada and had a successful Dental practice until his passing in 1916.



Despite the many strides that have been made to improve the lives of those living with sickle cell disease, the SCD community still faces numerous challenges. For instance:

 The average life expectancy of a person with SCD is still relatively young at 45 years of age.
 Presently there are four (4) medications available that have FDA approval to treat those with the disease.
 There is an overwhelming shortage of physicians (primarily hematologists) that treat or specialize in SCD which makes it difficult for patients to find specialized sickle cell disease care.
 To date there is no comprehensive model of care within federal agencies to help reduce the major healthcare complexities that SCD patients encounter.

 Though one of the oldest diseases existing, SCD receives a significantly disproportionate amount of funding for research and treatment compared to other diseases, particularly blood disorders and oncological disorders.
 Most importantly, 100 years since its discovery…there is still no universally available cure.

What Can I Do to Help?

You can:

- ☐ Get tested and know your trait status….
- ☐ Help raise awareness by talking about sickle cell disease to family, friends and associates.
- ☐ Become a blood donor and designate your donation to be reserved for a sickle cell patient.
- ☐ Volunteer with your local Sickle Cell Community organization for camps, programs, health fairs, and other activities.
- ☐ Donate to support better research.
- ☐ Participate in activities at your church that support sickle cell disease awareness!

For more information, go to www.sicklecelldisease.org. These are other useful websites: www.cdc.gov/ncbddd/sicklecell/; www.scinfo.org/

National Baptist Convention, USA, Incorporated
Dr. Jerry Young, President
Dr. Alvin Edwards, General Secretary
Dr. Alvin Love, Chair, Federal Faith-Based Initiative
Dr. Michael O. Minor, National Director, NBCUSA H.O.P.E. HHS Partnership
Dr. Genice Nelson, NBCUSA H.O.P.E. National Sickle Sabbath Sunday Chair

Sickle Cell Sabbath Sunday 2024 Worship Helps
Sunday, September 15, 2024

Responsive Reading

***Is there* no balm in Gilead,**
***Is there* no physician there?**
Why then is there no recovery
For the health of the daughter of my people? (Jeremiah 8:22, NKJV)

Now a certain woman had a flow of blood for twelve years, and had suffered many things from many physicians. She had spent all that she had and was no better, but rather grew worse.

When she heard about Jesus, she came behind Him in the crowd and touched His garment.

For she said, “If only I may touch His clothes, I shall be made well.”

***Immediately the fountain of her blood was dried up, and she felt in her body that she was healed of the affliction.* (Mark 5:25-29, NKJV)**

Blessed is he who considers the poor;
The LORD will deliver him in time of trouble.

The LORD will preserve him and keep him alive,
And he will be blessed on the earth;
You will not deliver him to the will of his enemies.

The LORD will strengthen him on his bed of illness;
You will sustain him on his sickbed. (Psalm 41:1-3, NKJV)

Litany

Adapted from Litany 95 "Health and Wellness" in the *Total Praise: Songs and Other Worship Resources for Every Generation* – Sunday School Publishing Board

Leader: We stand in Your presence, acknowledging You as our Creator and our all-knowing God.

People: We thank You for creating us in Your image and after Your own likeness.

Leader: We praise You, O God, for providing deliverance for our souls and healing for our bodies.

People: Help us to call upon You, for You are the one who blots out our sins and heals us from our diseases.

Leader: For the times when illnesses like sickle cell disease attack our bodies with seemingly no hope for a cure,

People: Help us, O God, to seek You for comfort when our bodies do not respond to the remedies that we employ. We will rest in the knowledge that one day we will have new bodies that are free of sickness and pain.

Leader: O God, You loved us so much that You gave Your Son. We now have eternal life, we have Your Spirit, we have Your Word, and we can enjoy healing that comes from You.

People: We thank You, Lord, for Your many blessings, and for those medical personnel who give of themselves in caring for the sick. Thank You for researchers who work relentlessly to find cures for illnesses that in the past were not available. Thank You for medical facilities all over the land that provide much-needed care.

Leader: It is You who has made us, and not we. Therefore, we will look to You as our Lord, our Savior, and our Healer.

All: We thank You, O Lord, that it is Your desire for us to enjoy good health, even as our souls prosper.

Sickle Sabbath Sunday Sermon Outlines

From "Hopeless" to Hopeful to Healing (Mark 5:25-29)

I. A "Hopeless" Situation (v. 25)
   a. Years of suffering
   b. Most likely single or divorcée in a male dominated society
II. "Hopeless" Results (v. 26)
   a. Medical treatment only made things worse
   b. Her financial resources were gone
III. Hopeful News (v. 27a)
   a. She heard about Jesus
   b. Jesus brings hope to a "hopeless" situation
IV. Hope Brings Action (vv. 27b-29)
   a. She pressed her way through the crowd
   b. She had faith in Jesus to be healed
   c. She was healed immediately
V. Hope Brings a Testimony (vv. 28-34)
   a. Jesus' asks a rhetorical question
   b. Jesus' disciples' negative response
   c. The woman testifies
   d. The woman is not only healed but blessed

Note: This woman did not have sickle cell disease.

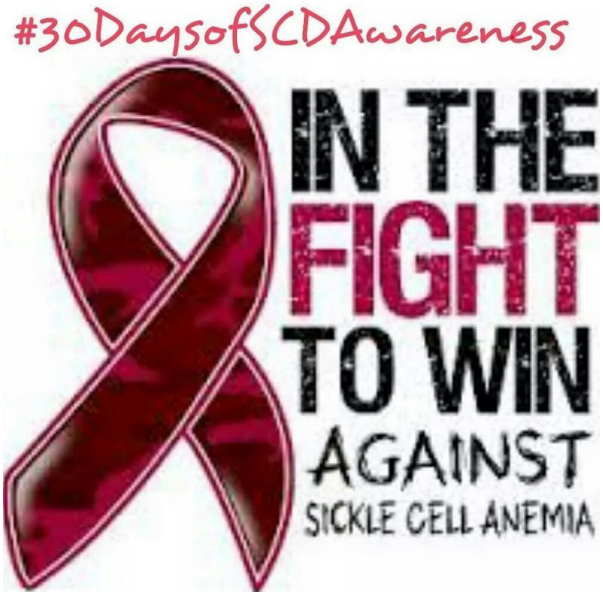
Why Do Our "Brooks" Dry Up? (1 Kings 17:1-9)

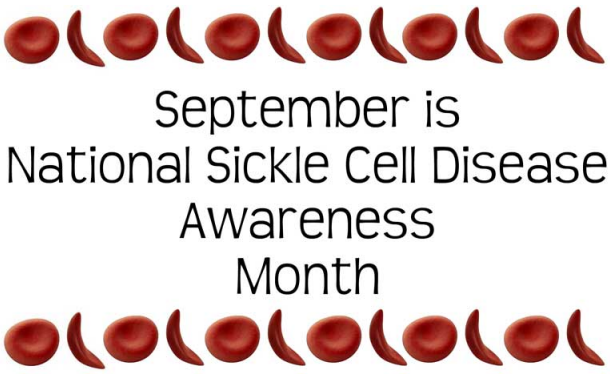
I. What Are Our "Spiritual" Brooks?
   a. Sources of blessings in our lives
   b. Our physical and spiritual families
   c. Our trust in God
II. Why Do Our Brooks Dry Up?
   a. The commonality of people: What can happen to you can happen to me
   b. The nature of living: We might fool "Mother Nature" but we can't out run "Father Time"
   c. The pressures of life: The more I pray the worse things *seem* to get
III. Why Are We Always Blessed in the End?
   a. The providential will of God: We can see to the corner, but God can see around the corner and beyond
   b. A whole life of suffering on earth is nothing compared to an eternal life of joy in heaven


Courage
Faith
Strength
Hope
SICKLE CELL ANEMIA AWARENESS


SCDAA
"Break The Sickle Cycle"®
September is National Sickle Cell Month


Someone I
LOVE
NEEDS A
CURE.
PLEASE SUPPORT
SICKLE CELL ANEMIA
AWARENESS


#30DaysofSCDAwareness
IN THE
FIGHT
TO WIN
AGAINST
SICKLE CELL ANEMIA


September is
National Sickle Cell Disease
Awareness
Month


Sickle Cell Disease
AWARENESS
...because it MATTERS

SICKLE CELL DISEASE

Sickle cell disease (SCD) is the most common inherited blood disorder in the United States. People with SCD are born with it. This disease gets its name from the abnormal crescent or "sickle" shape that some red blood cells develop. Because of this shape and other abnormalities, sickled red blood cells can block the flow of blood through the body and cause recurring episodes of pain, or pain crises.

CAUSES

SCD is caused by a change in hemoglobin, the protein in red blood cells that carries oxygen to all parts of the body. This type of hemoglobin is called hemoglobin S. When red blood cells with hemoglobin S go through the smallest blood vessels, called capillaries, some of the cells form into rigid strands and become sticky. These rigid, sticky strands often get stuck, clogging the capillaries. As a result, different parts of the body do not get the oxygen they need.

SIGNS AND SYMPTOMS

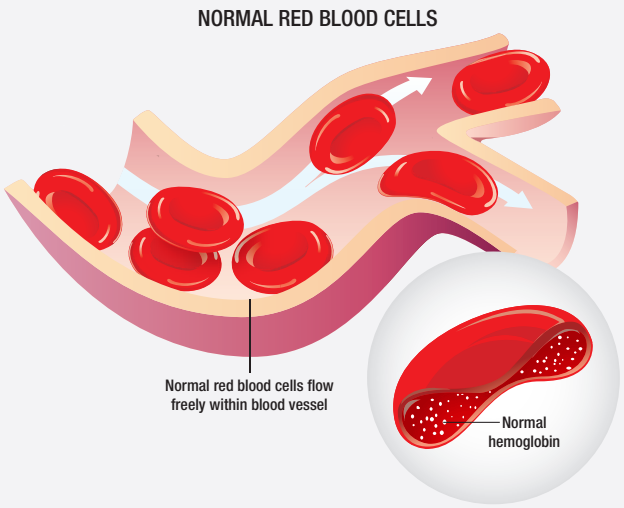
Most newborns who have SCD do not have symptoms until they are about 5 or 6 months old.

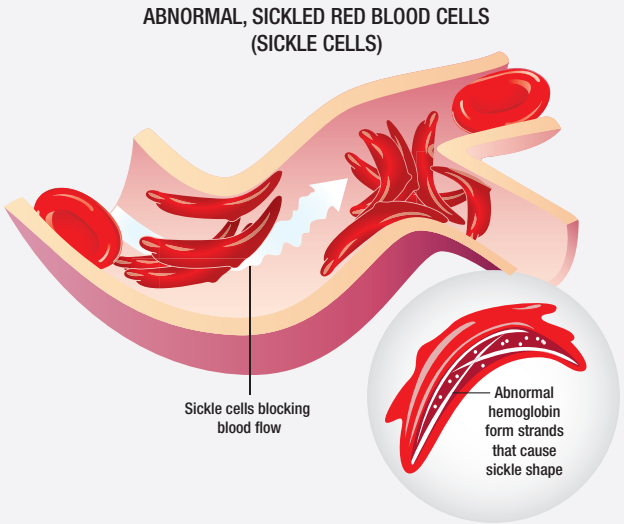
Symptoms can vary from person to person and can change over time. A person may or may not have symptoms depending on how SCD affects their health. Some people have symptoms once in a while; others have symptoms very often.

EARLY SIGNS AND SYMPTOMS

Early symptoms of SCD may include:

- Yellowing of the skin and whites of the eyes
- Fatigue or fussiness from anemia
- Painful swelling of the hands and feet





National Heart, Lung, and Blood Institute
Blood Diseases & Disorders Education Program
For more information, visit **sicklecell.nhlbi.nih.gov**
NIH Publication No. 22-HL-3058 July 2022

SICKLE CELL DISEASE

DIAGNOSIS

Doctors diagnose SCD using different screening blood tests. People who do not know whether they have the gene that makes sickle hemoglobin or another abnormal hemoglobin, such as SC, Sß thalassemia, and SE, can find out by having their blood tested. This helps them learn whether they carry a gene—or have the sickle cell trait—for an abnormal hemoglobin that they could pass on to a child. Every state in the United States, the District of Columbia, and the U.S. territories require that hospitals test babies for SCD as part of a newborn screening program.

TREATMENT

SCD is a lifelong illness. The good news is that several new medicines that will help ease the symptoms have been approved over the last few years. After a diagnosis, your doctor may recommend one of these medicines. In certain situations, blood transfusions may work better to manage complications, including chronic pain.

Currently, a bone marrow transplant is the only cure for SCD. But this is not for everyone. Many patients who have SCD either are too old for a transplant or do not have a donor who is a good genetic match. A patient needs a well-matched donor to have the best chance for a successful transplant.

Researchers are also exploring genetic therapies. Genetic therapies aim to repair a faulty gene or add a missing or new gene. These may help lead to new treatments or help cure SCD.

CLINICAL TRIALS

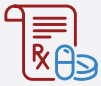
The National Heart, Lung, and Blood Institute continues to fund and conduct clinical trials for SCD. People who participate in clinical trials play an important role in helping to develop safe and effective new treatments and potential cures. Because of patients' contributions, researchers understand more about the causes of SCD and are developing ways to prevent and treat complications. Learn more about participating in a clinical trial.

REDUCING COMPLICATIONS

Here are a few ways to help relieve symptoms and reduce the chances of serious problems:

RECEIVE ROUTINE MEDICAL CARE.

STAY UP TO DATE ON VACCINES.



LEARN HOW TO MANAGE PAIN.

- When pain worsens, drink lots of fluids and take a nonsteroidal anti-inflammatory pain medicine such as ibuprofen. If you have kidney problems, doctors recommend acetaminophen instead.
- If you cannot control the pain at home, go to a SCD day hospital/outpatient unit or an emergency room to receive additional, stronger medicines and intravenous fluids.

ADOPT A HEALTHY LIFESTYLE.

- Exercise regularly.
- Choose heart-healthy foods.
- Drink water to avoid dehydration.
- Quit smoking. For free help and support, visit www.smokefree.gov.
- Get 7-9 hours of sleep a night.

National Heart, Lung, and Blood Institute

Blood Diseases & Disorders Education Program

For more information, visit **sicklecell.nhlbi.nih.gov**
NIH Publication No. 22-HL-3058 July 2022

FACTS AND STATS

WHAT?

Sickle cell disease (SCD) is an inherited blood disorder in which red blood cells may become sickle-shaped and harden. For a baby to be born with sickle cell disease, both parents must carry a sickle cell trait or genes for another hemoglobin like HbC, HbE or beta thalassemia. Sickle cell disease is not contagious, and there is no universal cure.

The Facts

- About **1 in 13** African Americans carry the sickle cell trait, and many do not know they have it.
- An estimated **100,000** people in the U.S. have SCD.
- Approximately **2,000** babies are born with SCD annually in the U.S.
- On average, diagnosis is made at **birth.**
- People of many ethnic **backgrounds** can have SCD.
- Latinos have the **second** most common incidence in the U.S.

The blockage of blood flow caused by sickled cells leads to complications including:

- Chronic severe and unpredictable pain
- Anemia
- Frequent infections
- Swelling in extremities
- Fatigue
- Delayed growth
- Vision problems/blindness
- Lung tissue damage
- Kidney disease
- Stroke
- Shortened life expectancy
- Damage to hip joint

WHERE?

- Sickle cell disease is a global health problem.

WHAT YOU CAN DO

- Donate blood to support transfusions.
- Advocate for better treatment, education and research.
- Educate others about sickle cell disease and sickle cell trait.
- Get tested for sickle cell trait if you are of African descent and do not know your status.
- Support SCDAA as we search for a universal cure.

5 FACTS YOU SHOULD KNOW ABOUT SICKLE CELL DISEASE

A child gets sickle cell disease (SCD) when he or she receives two sickle cell genes*—one from each parent.

A child who inherits only one sickle cell gene has sickle cell trait (SCT). If both parents have either SCD or SCT, it is important for them to discuss this information with each other and with a doctor when making decisions about family planning.

*Genes, which are passed down from a parent to child, are instructions in each of our cells that determine a person's traits such as eye color, blood type, and risk of disease.

SCD has many faces.

The disease affects millions of people worldwide and is especially common among people who come from and whose ancestors come from the following regions highlighted in red:

SCD can be cured for certain patients.

Bone marrow transplants (BMTs) and newly developed gene therapies are potentially curative treatment options for some patients. A BMT, which involves collecting healthy cells from a donor's bone marrow and transferring them into a patient, can cure SCD. However, it may not be the best choice for all patients because it comes with serious risk. A BMT expert can advise patients about whether it is a good choice for them.

Gene therapies for the treatment of SCD are now approved for use in patients 12 years and older. While these therapies mark major advances in the treatment of SCD, they are so new more data are needed to understand their impact on the patient and their chance of recovery.

Anemia is a common effect of SCD, but it can be treated.

In someone with SCD, red blood cells die early and not enough are left to carry oxygen throughout the body, causing anemia. Infection or enlargement of the spleen, an organ that stores red blood cells, may make anemia worse. Blood transfusions are used to treat severe anemia.

A person with SCD can live a long and high quality life.

More than **95%** of newborns with SCD in the United States will live to be adults. People with SCD can lower their chances of difficulties from the disease and enjoy many normal activities by

✓ Getting regular checkups with their doctor.

✓ Following treatments prescribed by their doctor, such as taking medication called hydroxyurea.

✓ Preventing infections by taking simple steps including washing their hands.

✓ Practicing healthy habits like drinking 8 to 10 glasses of water per day and eating healthy food.

For more information about SCD, visit: **www.cdc.gov/ncbddd/sicklecell**

CDC U.S. CENTERS FOR DISEASE CONTROL AND PREVENTION

SICKLE CELL DISEASE

STAYING WELL

HOW?

People with SCD can live full lives by being proactive in their care. Here are some tips to help you or someone you know with SCD stay as healthy as possible.

Staying healthy with sickle cell disease involves:

- Pain management
- Preventing infections from common illnesses such as the flu
- Self-care including eating well, exercising and staying hydrated
- Medications to reduce the severity of sickle cell disease
- Regular preventive care including vaccinations and health and dental check ups
- Quality medical care from doctors and nurses who are educated about SCD
- Building a support system of friends and family
- Connecting with a patient support group or a community-based SCD organization for information and assistance

TREATMENT OPTIONS

- Over-the-counter pain relievers are commonly used to treat chronic pain.
- Hydroxyurea (Siklos, Droxia) and Oxbryta (Voxelotor) are prescribed medications that can decrease some complications of SCD.
- Endari (Glutamine) and Adakveo (Crizanlizumab) are prescribed medications that can reduce the number of sickle cell pain crises.
- Blood transfusions can help relieve symptoms of SCD and potentially prevent complications.

SCREENING TESTS

- Newborn screening can help identify SCD and expedite early management and treatment.
- Adult screening can help identify if someone has SCD or is a carrier for the SCD trait.
- Sickle stroke screening, aslo known as Transcranial Doppler (TCD) screening, can help identify people with brain abnormalities as a result of SCD who are at high risk for ischemic stroke and brain injury.
- Organ screening can help identify damage to organs such as the kidney, eye and heart in adults.

THE SEARCH FOR A CURE...

Bone marrow (stem cell) transplants can, in some cases, cure sickle cell disease, but not all individuals are eligible for this procedure, and there are associated risks. Read more about this NIH initiative: **www.curesickle.org**

This information is for educational purposes only and does not serve as medical advice or as an endorsement by SCDAA. Talk to your doctor about the screenings and treatments that may be right for you.

Healthy Living With Sickle Cell Disease

If you have sickle cell disease (SCD), you can live a full life and enjoy the things that most other people do. Follow these tips to support your mental and physical health, manage your condition, and stay as healthy as possible.

See Your Healthcare Team Regularly

Work closely with your healthcare team to develop your unique care plan. Try to establish strong relationships with your hematologist and other specialists, and keep in touch with them as often as needed. Routine checkups can help prevent serious problems. Your healthcare team can offer specific guidance, prescribe medications and treatment options, and address any concerns or questions. Most people with SCD should see their provider every 3 to 12 months. It's also important to get vaccines as directed by your healthcare team, including a flu shot each year, and a COVID-19 vaccine, as recommended.

Manage Your Blood Pressure

People with SCD are at high risk for complications, including stroke and kidney disease. These complications can be worsened by high blood pressure (hypertension), which often adds to the workload of the heart and blood vessels. Ask your healthcare team to check your blood pressure at each visit, and ask what your numbers mean for you — and how to control your blood pressure if it's consistently higher than 130/80 mm Hg.

Take Care of Your Mental Health

Children and adults with SCD may experience pain, anxiety, and depression. Talk to your friends and family about how you are feeling. Your healthcare team can refer you to a mental health professional to develop healthy pain-coping strategies, address negative thoughts, and improve overall well-being. Supportive counseling and, sometimes, antidepressant medicines may help.

Adopt a Healthy Lifestyle

Maintaining a healthy lifestyle is important for overall well-being and managing SCD.

- Be physically active
- Choose heart-healthy foods
- Drink 8–10 glasses of water daily
- Limit alcohol
- Quit smoking or vaping
- Wash hands often to help prevent infections
- Take care of your mental health
- Aim for 7–9 hours of sleep each night

Healthy Living With Sickle Cell Disease

Learn What to Do in a Pain Crisis

When an acute crisis is just starting, drink lots of fluids and take a nonsteroidal anti-inflammatory (NSAID) pain medicine, such as ibuprofen. If you have kidney problems, acetaminophen is often preferred. If you cannot manage the pain at home, go to a day hospital or outpatient unit or an emergency department to receive additional stronger medicines and IV fluids. You may need to be admitted to the hospital to fully control an acute pain crisis.

Know When to Seek Help

Have a plan to get help quickly if there is a problem. Keep a copy of your health records (electronic or print) so you can bring them to a healthcare facility as needed.

Call 9-1-1 or seek emergency room care right away if you have:

- Fever above 101° F
- Difficulty breathing
- Chest pain
- Abdominal (belly) swelling
- Sudden vision problems
- Severe headache
- Sudden weakness or loss of feeling and movement
- Seizure
- Painful erection of the penis that lasts more than 4 hours
- Pain anywhere in the body that will not go away with treatment

By adopting a healthy lifestyle, you can help take control of your well-being. For more information on a healthy lifestyle with SCD, visit sicklecell.nhlbi.nih.gov

"I drink lots of fluids, eat a healthy diet, take my medications, even if I'm feeling good, and make every doctor's appointment," says Yaw Lokko, patient advocate and clinical research trial participant. And to minimize pain triggers, "I try to avoid the heat, avoid the cold. It's not always easy, but it's good to pay attention."

National Heart, Lung, and Blood Institute

Blood Diseases & Disorders Education Program

NIH Publication No. 23-HL-8211 August 2023

Managing Pain With Sickle Cell Disease

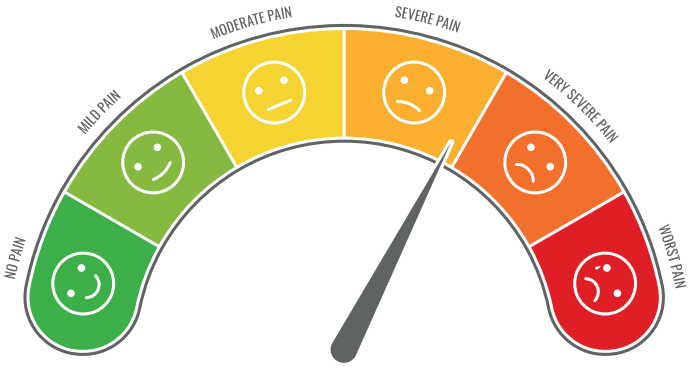
People with sickle cell disease (SCD) may experience pain in different ways. The sickled cells that give the disease its name can lead to pain throughout the body and serious damage to organs, such as the heart and kidneys. If you have SCD, see your healthcare team regularly to help prevent pain, complications, and serious problems. Work with your healthcare team to create a pain management plan that makes sense for you.

Common Types of Pain

- **An acute or severe pain crisis** can happen without warning when sickle cells block blood flow. People describe this pain as sharp, intense, stabbing, or throbbing.
- **Pain from organ damage** may occur when sickled cells affect your heart, kidneys, spleen, or other body parts.
- **Chronic or long-term pain** is also common, but it can be hard to describe. It is usually different from crisis pain or the pain that results from organ damage.
- **Joint problems and pain** may develop if there is sickling that lowers oxygen flow. This can occur in the hip bones and, less commonly, the shoulder joints, knees, and ankles.
- **Priapism** is an unwanted and sometimes prolonged painful erection. This happens when blood flow out of the erect penis is blocked by sickled cells.

Tips to Manage Pain

- Most patients can sense when an acute crisis is just starting. Drink lots of fluids and take a nonsteroidal anti-inflammatory (NSAID) pain medicine, such as ibuprofen. If you have kidney problems, acetaminophen is often preferred.
- If you cannot manage the pain at home, go to a day hospital or outpatient unit or an emergency department to receive additional, stronger medicines and IV fluids. Your healthcare team may prescribe stronger medicines called opioids for severe pain.
- In the event of priapism, seek medical care immediately if symptoms last more than four hours or if it happens more than once within a 24-hour period.
- Keep a copy of your pain management plan (print or electronic) when you seek emergency care.



"If I feel pain within a day or two and don't do something about it, I'll be in a full-blown sickle cell crisis."

That internal barometer has helped Ebow H-Smith, healthcare professional and sickle cell disease advocate, manage pain as an adult.

Managing Pain With Sickle Cell Disease

Tips to Manage Pain *(Continued)*

- Other methods for managing pain include physical therapy, acupuncture, using a heating pad, massage, yoga, or guided audiovisual relaxation.
- Talk to your healthcare team about when to seek emergency care.

Prevent Problems Over Your or Your Child's Lifetime

- **Avoid situations that may set off a crisis.** Extreme heat or cold, as well as sudden changes in temperature, are often triggers. When going swimming, ease into the water rather than jumping right in.
- **Do not travel in an aircraft cabin that is unpressurized.**
- **If you experience priapism**, you may be able to relieve your symptoms by doing light exercise, taking a warm bath or shower, emptying your bladder by urinating, drinking more fluids, and taking medicine recommended by your healthcare provider.
- **If your child attends day care, preschool, or school, speak to their teacher about the disease.** Teachers need to know what symptoms to watch for and how to accommodate your child.
- **Ask your healthcare team about medications that may control SCD and help reduce the risk of pain crises.**

Questions to Ask Your Healthcare Team About Treatments

- Which treatments are best for me?
- How is this treatment administered?
- How often do I need this treatment?
- What are the side effects?

With help from your healthcare team, you can create a plan to manage your pain and prevent serious problems. Learn more at **sicklecell.nhlbi.nih.gov**

National Heart, Lung, and Blood Institute

Blood Diseases & Disorders Education Program

NIH Publication No. 23-HL-8212 August 2023

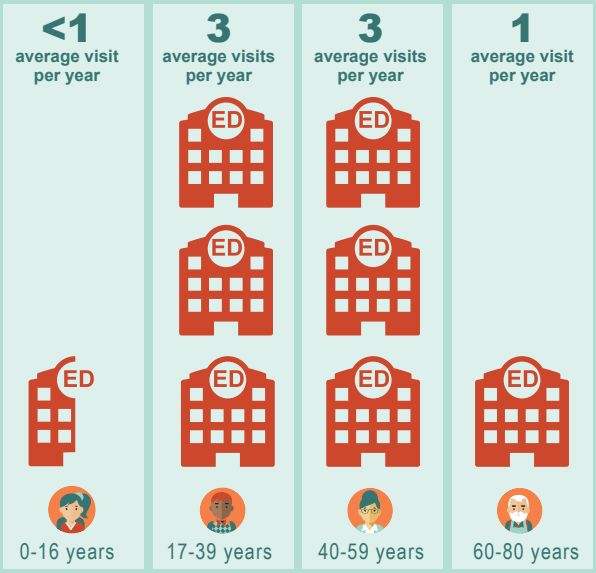
DO YOU USE THE EMERGENCY DEPARTMENT FOR CARE OF SICKLE CELL DISEASE?

WHAT TO KNOW BEFORE YOU GO.

Children and adults with sickle cell disease (SCD) often require care in the emergency department (ED) of hospitals or clinics for health issues related to SCD. The ED may be your only option for health care when symptoms, such as pain crises, cannot be managed at home or when you do not have access to a healthcare provider who specializes in treating SCD.

The Sickle Cell Data Collection (SCDC) program found that in California, people with SCD seek care in the ED an average of three times a year from their late teens to their late 50s. *Excruciating pain, known as a sickle cell crisis, is the most common reason for these ED visits.*

Emergency Department (ED) Visits Among People with Sickle Cell in California, 2005-2014



Tips for receiving better care in the ED

Before you get sick or have a pain crisis, work with your regular doctor to

- Make sure that information in your electronic medical record (EMR) is updated, including your medical history and current pain medicines.

- Create a pain management plan and make sure it is entered into your EMR. Keep with you a printed copy of the plan and a list of all your medicines.

When you go to the ED

- Tell the ED staff right away that you have SCD.
- Share openly with your ED nurse and doctor

- Your medical history, including a list of your medicines.
- Your pain management plan. Ask the ED nurse or doctor to look up your plan in your EMR or share a printed copy.
- Your regular doctor's contact information. If the ED nurse or doctor has concerns about your pain management, ask the ED staff to call your regular doctor.

PROTECTING PEOPLE

CDC's National Center on Birth Defects and Developmental Disabilities is committed to protecting people and preventing complications of blood disorders. Learn more about CDC's work to help people with SCD here: www.cdc.gov/ncbddd/sicklecell

U.S. Department of Health and Human Services
Centers for Disease Control and Prevention

SCDC
Sickle Cell Data Collection

Hydroxyurea Use for Sickle Cell Disease

Hydroxyurea is an oral medicine that can help reduce sickle cell disease (SCD) complications.

People who take hydroxyurea must use it regularly as prescribed and receive consistent medical care. Ask your healthcare provider about the potential benefits and risks, and whether hydroxyurea is right for you.

Facts About Hydroxyurea

It has been a standard treatment for SCD for more than 30 years.
The Food and Drug Administration (FDA) approved hydroxyurea for severe SCD in 1998 for adults and in 2017 for children.

It can reduce the likelihood of a pain crisis.
Hydroxyurea has been shown to reduce, by about half, the number of painful events a person with SCD may experience.

It has been shown to improve anemia and reduce the need for blood transfusions and hospital admissions.
One way to help reduce fatigue from anemia in people living with SCD is by increasing hemoglobin levels. This provides more oxygen to the body, giving the person with SCD increased energy. Studies have shown hydroxyurea is one of several drugs that does this.

Most experts recommend daily use of hydroxyurea for children and adults with sickle cell disease.

It can reduce the number of episodes of acute chest syndrome, a medical emergency.
By increasing hemoglobin, hydroxyurea can potentially prevent the blockage of blood flow to the lungs.

Hydroxyurea Use for Sickle Cell Disease

It may reduce the risk of stroke in children with SCD.
SCD reduces blood flow to the brain, which can cause a stroke, particularly in children.

It is not appropriate for anyone who is pregnant.
If you are thinking of becoming pregnant, talk to your doctor about when to stop taking hydroxyurea.

Questions to Ask Your Healthcare Provider

- How does hydroxyurea treat SCD?
- What are the possible short- and long-term side effects for me?
- How long would I need to take it?
- How many pills would I need to take every day?
- How often would I need to see my doctor when taking hydroxyurea?
- Are there other treatments I should consider?

SCD treatments, including hydroxyurea, may reduce symptoms and improve quality of life. Work with your healthcare provider to find what works best for you.

Learn more about available SCD treatments at sicklecell.nhlbi.nih.gov

National Heart, Lung, and Blood Institute
Blood Diseases & Disorders Education Program
NIH Publication No. 24-HL-8219 February 2024

Crizanlizumab

TREATMENT FOR SICKLE CELL DISEASE

Crizanlizumab (pronounced "criz-an-liz-u-mab"), or **ADAKVEO®** is a treatment for sickle cell disease (SCD). It may help people ages 16 years and older who live with all types of SCD (If you don't know the type of SCD you or the person receiving care has, ask your doctor). Share the information in this fact sheet with your health care team to decide whether this drug might be a good choice for you or the person receiving care. You may be able to take this drug even if you are currently taking hydroxyurea, but consult with your health care team. Crizanlizumab has not been studied in women who are pregnant or breastfeeding.

How crizanlizumab works

- For individuals living with SCD, red blood cells are distressed, which makes them stiff, sticky, and sickled-shaped. This stickiness makes blood cells clump and block blood flow in a process called vaso-occlusion, which causes severe SCD pain crises.
- Crizanlizumab works by slowing or stopping red blood cells and other blood cells, like white blood cells and platelets, from sticking to blood vessel walls.
- Crizanlizumab acts like an oil slick, spreading over and between cells, which helps the blood flow and decreases the number of pain crises.

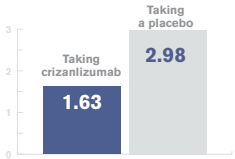
How crizanlizumab is given

- Crizanlizumab may be administered monthly and is given intravenously (IV), or into the vein, over a period of 30 minutes every four weeks. Talk to your doctor if you have a history of problems with IV treatment.
- This drug may be administered in many settings depending on your health care team and your insurance provider's policies. You can take this drug at your doctor's office or an infusion center, or an outpatient day hospital.
- Crizanlizumab is for long-term use. Talk to your health care team about how long will be right for you or the person receiving care.

Research study of crizanlizumab

- During a clinical research study known as the SUSTAIN study, participants ages 16 and over living with SCD who took crizanlizumab reported fewer pain crises than those on placebo (a harmless substance that looks like the treatment but is specifically designed to do nothing).
- Over the course of a year, the participants taking crizanlizumab reported 45% fewer pain crises: participants on crizanlizumab had an average 1.63 pain crises while those on the placebo averaged 2.98 crises.
- Some of the side effects of crizanlizumab (occurring in at least 1 out of 10 people) include reactions to the IV, nausea, fever, back pain, and joint pain. These reactions also occurred in people taking the placebo but in only half as many people.

Average number of crises events after 48 weeks



People experiencing side effects on crizanlizumab

People experiencing side effects taking a placebo

Talk to your health care team about…

- The possible benefits and risks of crizanlizumab for you or the person receiving care
- Whether other health conditions or taking other medications might limit taking this drug
- Past reactions to drugs that may give clues about whether you or the person receiving care could have an allergic reaction to crizanlizumab
- Other treatment options
- How long you or the person receiving care should take crizanlizumab

About This Fact Sheet: This fact sheet has been prepared by the Sickle Cell Disease Coalition and provides general information about crizanlizumab, which is one of the few FDA-approved therapies to treat sickle cell disease. This fact sheet is purely an informational resource and is intended to facilitate discussion with your physician. It does not provide medical advice and is not intended to substitute for consultation with a medical professional. People with sickle cell disease should talk to their doctor before making any changes to their treatment.

For more information about SCD go to *www.scdcoalition.org*

SICKLE CELL DISEASE COALITION

L-Glutamine

TREATMENT FOR SICKLE CELL DISEASE

L-glutamine (pronounced "L-gloo-ta-meen,"), or **ENDARI®** is a treatment for sickle cell disease (SCD) that may help patients age five years and older (If you don't know the type of SCD you or the person receiving care has, ask your doctor). Share the information in this fact sheet with your health care team to decide whether this drug might be a good choice for you or the person receiving care. You may be able to take this drug even if you are currently taking hydroxyurea, but consult with your health care team. L-glutamine has not been studied in women who are pregnant or breastfeeding.

How L-glutamine works

- L-glutamine is an amino acid. The body naturally produces amino acids to help prevent and fight against infection, injury, and stress. However, people with SCD might need additional L-glutamine because they experience more pain and infection than what the body can handle on its own.
- For individuals living with SCD, red blood cells are distressed, which makes them stiff, sticky, and sickled-shaped. This causes damage that blocks blood flow in a process called vaso-occlusion, which can cause pain crises. L-glutamine helps stop or slow down this damage and works to lower the number of pain crises.

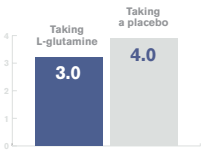
How to take L-glutamine

- L-glutamine is taken in powder form and is intended for long-term use.
- Each dose normally is mixed in eight ounces of cold or room temperature beverage or four to six ounces of food.

Research study of L-glutamine

- In a study looking at the effectiveness of L-glutamine, researchers found that participants assigned to take L-glutamine had fewer pain crises than those on the placebo (a harmless substance that looks like the treatment but is specifically designed to do nothing).
- After 48 weeks, the L-glutamine group had fewer crisis events than the placebo group (median of 3 instead of 4 events).
- People who took L-glutamine went to the hospital less often than the other group (median of 2 instead of 3 hospitalizations).
- L-glutamine may help decrease other sickle cell complications. Please discuss further with your health care team.
- Some of the side effects of L-glutamine (occurring in at least one in 10 people – same as placebo) include constipation, nausea, headache, stomach pain, cough, leg or arm pain, back pain, and chest pain.

Average number of crises events after 48 weeks



People experiencing side effects on L-glutamine or placebo

Talk to your health care team about…

- The possible benefits and risks of L-glutamine for you or your child
- Whether other health conditions or taking other medications might limit taking L-glutamine
- Other treatment options
- How long you or the person receiving care should take L-glutamine.

About This Fact Sheet: This fact sheet has been prepared by the Sickle Cell Disease Coalition and provides general information about L-glutamine, which is one of the few FDA-approved therapies to treat sickle cell disease. This fact sheet is purely an informational resource and is intended to facilitate discussion with your physician. It does not provide medical advice and is not intended to substitute for consultation with a medical professional. People with sickle cell disease should talk to their doctor before making any changes to their treatment.

For more information about SCD go to *www.scdcoalition.org*

SICKLE CELL DISEASE COALITION

Voxelotor

TREATMENT FOR SICKLE CELL DISEASE

Voxelotor (pronounced "vox-EL-o-tor"), or **OXBRYTA®** is a treatment for sickle cell disease (SCD) that may help people aged 12 years and older (If you don't know the type of SCD you or the person receiving care has, ask your doctor). Share the information in this fact sheet with your health care team to decide whether this drug might be a good choice for you or the person receiving care. You may be able to take this drug even if you are currently taking hydroxyurea, but consult with your health care team. Voxelotor has not been studied in women who are pregnant or breastfeeding.

How voxelotor works

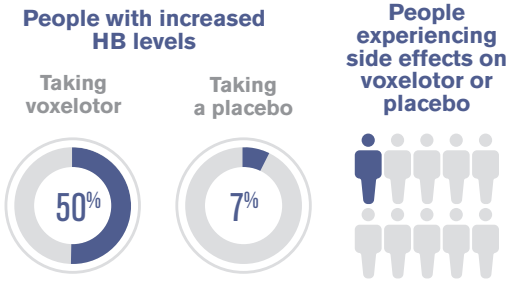
- Healthy red blood cells contain hemoglobin (Hb), which carries oxygen to all parts of your body.
- For individuals living with SCD, red blood cells lose their round shape and become sickled, or crescent-shaped.
- The sickled red blood cells break apart in a process called hemolysis.
- Voxelotor decreases hemolysis and strengthens hemoglobin's carrying of oxygen.

How to take voxelotor

- Voxelotor is given as a pill. The typical dose is three pills (1,500 mg) by mouth daily, with or without food but patients should consult with their health care team.
- Tablets should not be crushed.
- Voxelotor is for long-term use. Talk to your health care team about how long will be right for you or the person receiving care.

Research study of voxelotor

- During a clinical study known as the HOPE study, 50% of participants between the ages of 12 and 64 years were found with increased Hb levels (i.e. in two to four weeks), whereas only 7% of those given the placebo (a harmless substance that looks like the treatment but is specifically designed to do nothing) had increased Hb levels.
- After 24 weeks, the group given voxelotor also showed fewer hemolysis events than the placebo group.
- Side effects of voxelotor (occurring in at least one in 10 people - same as placebo) include headache, diarrhea, stomach pain, nausea, tiredness, rash, or fever.



Talk to your health care team about…

- The possible benefits and risks of voxelotor for you or the person receiving care
- Whether other health conditions or taking other medications might affect taking voxelotor
- Past reactions to drugs that may give clues about whether you or your child could have an allergic reaction to voxelotor
- Other treatment options
- How long you or the person receiving care should take voxelotor

About This Fact Sheet: This fact sheet has been prepared by the Sickle Cell Disease Coalition and provides general information about voxelotor, which is one of the few FDA-approved therapies to treat sickle cell disease. This fact sheet is purely an informational resource and is intended to facilitate discussion with your physician. It does not provide medical advice and is not intended to substitute for consultation with a medical professional. People with sickle cell disease should talk to their doctor before making any changes to their treatment.

For more information about SCD go to *www.scdcoalition.org*

SICKLE CELL DISEASE COALITION

AMERICAN SOCIETY OF HEMATOLOGY
2021 L Street, NW, Suite 900, Washington, DC 20036 ph 202.776.0544 fax 202.776.0545 e-mail ASH@hematology.org

Sickle Cell Disease Fact Sheet

REQUEST: Cosponsor legislation to enhance federal government activities in sickle cell disease SCD research, training and services.

Sickle Cell Disease (SCD) and Sickle Cell Trait (SCT)

SCD is an inherited, lifelong disorder affecting nearly 100,000 Americans. Individuals with the disease produce abnormal hemoglobin which results in their red blood cells becoming rigid and sickle-shaped and causing them to get stuck in blood vessels and block blood and oxygen flow to the body. SCD complications include severe pain, stroke, acute chest syndrome (a condition that lowers the level of oxygen in the blood), organ damage, and in some cases premature death.

Sickle cell trait (SCT) is **not** a disease. Having SCT simply means that a person carries a single gene for sickle cell disease (SCD) and can pass this gene along to their children. People with SCT usually do not have any of the symptoms of SCD and live a normal life.

State of SCD Care and Research

- Although the molecular basis of SCD was established many decades ago, it has been challenging to translate this knowledge into the development of novel targeted therapies.
- New approaches in managing this disease have improved diagnosis and supportive care over the last few decades, but many patients still have severe complications to overcome.
- SCD patients encounter major issues accessing high quality care. Because SCD is a complex illness that affects multiple organ systems, few physicians specialize in SCD or can assume primary responsibility for a patient's care. Coordinated treatment continued across multiple settings is essential for patients to receive adequate care.
- With funding from the CDC Foundation, the Centers for Disease Control and Prevention (CDC) has established a population-based surveillance system to collect and analyze longitudinal data about people living in the U.S. with SCD. Due to limited funding, however, implementation of the program has occurred only in two states – California and Georgia (approximately 10% of the U.S. SCD population). Data is being collected from multiple sources (newborn screening programs and Medicaid) in order to create individual healthcare utilizations profiles.

Steps Needed to Bridge the Gap in Care and Research

There is a critical need to improve outcomes for patients suffering with this disease. Expanded surveillance is necessary to improve understanding of the health outcomes and health care system utilization patterns of people with SCD, to increase evidence for public health programs, and to establish cost-effective practices to improve and extend the lives of people with SCD. Given the exciting and promising new SCD research announced at the December 2016 American Society of Hematology (ASH) Annual Meeting, now is the time to make the investment to ensure that patients have access to state of state-of-the-art clinical care.

Strengthening and expanding current efforts will help enable individuals living with this disease receive adequate care and treatment. Legislation has been introduced in the House by Representatives Danny Davis (D-IL) and Michael Burgess (R-TX) – H.R. 2410, the Sickle Cell Disease Research, Surveillance, Prevention, and Treatment Act of 2017 – that seeks to enhance federal government activities in sickle cell disease SCD research, training and services. Specifically, the legislation reauthorizes SCD prevention and treatment grants awarded by the Health Resources and Service Administration (HRSA), authorizes the CDC to award SCD surveillance grants to states, and authorizes research to expand the understanding of the cause and find a cure for SCD. Bipartisan companion legislation is expected to soon be introduced in the Senate.

****

The American Society of Hematology (ASH) represents more than 17,000 physicians, researcher, and medical trainees committed to the study and treatment of blood and blood-related diseases. ASH members include clinicians who specialize in treating children and adults with SCD and researchers who investigate the causes and potential treatments of SCD manifestations. ASH is committed to addressing the burden of SCD and recently released a *Call to Action on SCD* along with other stakeholders, founded the Sickle Cell Disease Coalition, and a public relations campaign. ASH's *State of SCD 2016 Report* and Report Card identified outlines the most pressing areas of need and provides a blueprint to advance these actions related to access to care, research and clinical trials, and global issues in sickle cell disease (SCD). For more information about the report, the report card and the new Sickle Cell Disease Coalition visit *www.scdcoalition.org* and for more information on SCD visit ASH's website (*www.hematology.org/SCD*).