Uriel E. Owens Sickle Cell Disease Association of the Midwest
444 Minnesota Ave. Suite 300
Kansas City, Kansas 66101
913-735-2622

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House Heatlh and Human Services

Representative Brenda Landwehr, Chairman

From: Kevin Wake

Good afternoon and thank you Madam Chairwoman and the members of the committee. I am Kevin Wake and I live in Kansas City. I am the President of the Uriel Owens Sickle Cell Disease Association of the Midwest and I am a 54 year old sickle cell disease patient and I have had 3 strokes as an adult related to my sickle cell disease. Our Association has provided resources such as emergency financial assistance, public education and awareness, advocacy, and educational scholarships for the sickle cell community for the past 42 years. I appreciative to speak briefly about sickle cell disease and the positives that HB2338 will do for both the sickle cell community and healthcare providers.

Sickle cell disease is a genetic blood disorder that affects over 100,000 patients in the US and over 700 Kansans. 1 in 13 African Americans are estimated to be trait carriers of this disease, meaning that the potential to pass this on to children is great. Sickle cell causes a person's red blood cells to be shaped like a crescent moon or a banana instead of being round. These cells also are rigid, sticky, and only live for about 10-20 days instead of the typical 120-day lifespan of normal cell. Sickle cells can't carry the appropriate oxygen throughout the body. The shape of these cells also causes the cells to not flow nicely throughout the body. These cells will often coagulate in the body called occlusions and cause the hallmark symptom that patients experience, and that is severe pain. This pain often requires opioid medication to control. Patients can also have organ and tissue damage wherever blood flows, which is everywhere throughout the body.

I was diagnosed in 1969 after my older brother passed away at the age of 2 ½ and sickle cell was discovered on his autopsy. I was 9 months old. My younger brother was born 2 years after my diagnosis, and he too was diagnosed as having sickle cell disease at birth.

I feel extremely blessed to have had parents that were supportive of our disease and did everything to ensure that we were cared for. My dad was a dairy farmer in Leavenworth and

my mom was a nurse, who put her career on hold to dedicate herself to being a fulltime caregiver. Growing up, we had to travel over an hour each way for appointments and hospitalizations. I now realize that the support that we had growing up is not the case for many families impacted with SCD. Mom encouraged us to live as normally as possible. To this day, I remember the checkup when mom asked about what steps should she take to prepare us for college and she was told, with us in the room, that she should not push us to go to college and let us live and enjoy our lives as best we can since most children with SCD don't live past 25.

Luckily, mom didn't accept that, and we both attended college and I had a 23 year career in the pharmaceutical industry before taking an early retirement due to health complications in 2016. Like many SCD patients, I experienced racial bias throughout our healthcare system. I waited for over 5 hours before getting treatment for a pain crisis in an emergency room in Dallas TX. I had to prove to the staff that I was a SCD patient, that my pain was real, and that my knowledge of what pain medication worked best for my pain didn't mean that I was a drug seeker.

In 1999, when I had my first sickle cell related stroke, I was working in Chicago and the EMT first reported my transfer to the hospital as a 'black male in his 30's...probable drug overdose or intoxication.' I was unable to tell them differently because the stroke left me unable to speak. Thankfully, I was able to write with my non dominate hand three words...sickle cell stroke and I was able to convince a nurse and doctor to access me differently and that probably saved my live. You can't simply look at a person and tell that they have sickle cell, so unfortunately, patients often face the stigma of being drug seeking in the ER. That bias within healthcare for SCD patient care continues to exist today. Adult sickle cell care clinics are few and far between, there is an overall lack of providers proficient in treating SCD, current treatment standards of care are rarely practiced on a routine basis, and the list goes on. I feel that a lot of this stems from the lack of awareness and education on a disease that has been known about for over a century.

Since my retirement, I have fallen into the role of patient advocacy. I often refer to sickle cell as the forgotten disease & the reason I say that is for a disease that was first discovered in the US in 1910, there have been few advances in terms of treatments. The first disease modifying agent approved by the FDA was hydroxyurea in 1988. The second approve treatment didn't come along until 2017, followed by two additional drug treatments in 2019.

Over 95% of children with sickle cell now survive into adulthood. This a direct reflection on improved treatments for kids and coordinated care models for peds, where all services are managed and coordinated through the pediatric centers. Sickle cell disease is one of the 34 for conditions screened for here in KS and is now on all the states newborn screening panels in the US. This means that patients can be identified at birth and be referred for treatment much earlier now.

Unfortunately, we see adult patients often get lost in the health system as they transition from pediatric care to adult care. Patients have few choices of SCD specific centers, there are

a lack of healthcare provers that specialize in the management of SCD, and patients must navigate the healthcare system on their own, and they take on this stressor with the many other stressors of being an adult, holding a job, paying rent, etc. Patients become reactive instead of proactive in their care. We see many patients treating their disease through the ER's, again where they often face bias, a lack of knowledge of SCD and few standardized treatment practices. This also leads to an overall increase in healthcare treatment costs due to treating complications instead of embracing preventative care for these patients.

We as a community are hearing a lot about gene therapies and the possibly cure soon. Along with these scientific advances, we need to raise awareness of the disease, seek out ways to improve patient access to care, especially for patients as they transition from pediatrics to adult care, and promote healthcare coordination. This will ultimately reduce ER department visits and inpatient hospital costs. HB2338 will be a big step in the right direction in addressing some of the many concerns that I have.

First, this legislation will establish the 3rd full week of September as sickle cell awareness week in the state. This will create awareness for this disease across the state. As I mentioned before, awareness is still greatly needed for disease awareness and education, but it is also needed to for the thousands of Kansans that carry the trait status so that they are aware of the risks prior to having children. MO passed legislation last year designating the same week of September as Sickle Cell Awareness week and I can vision a great collaboration between the two states in driving awareness here in the Midwest.

The second part of HB2338 will help be proactive in reviewing available treatments for sickle cell and better prepare the state in providing those treatments for patients. Again, proactive care and the use of some of these treatment developments will benefit patient care, and even can decrease the overall cost burdens that this disease has.

For these reasons, I ask for full support of HB2338 both to improve the lives of Kansans living with sickle cell and their families and to raise awareness so that those with the trait can make informed decisions prior to having children. This bill will also better prepare the State on available and late state treatment options to help improve the lives of Kansans as well.

Thank you for your time today and I welcome any questions that you may have.

Kevin Wake

President

"Help Break the Sickle Cycle"
fax: 913-371-8522 info@sicklecellmidest.org www.sicklecellmidwest.org

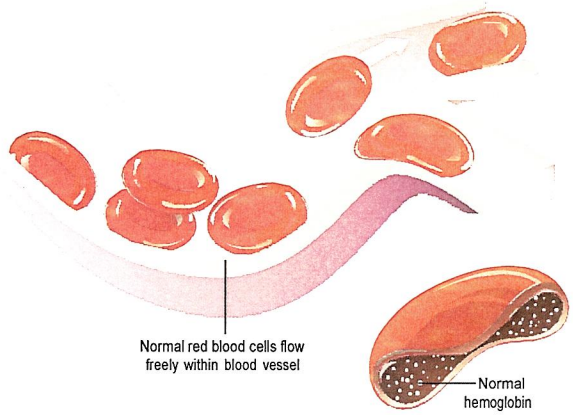
SICKLE CELL DISEASE

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.

NORMAL RED BLOOD CELLS

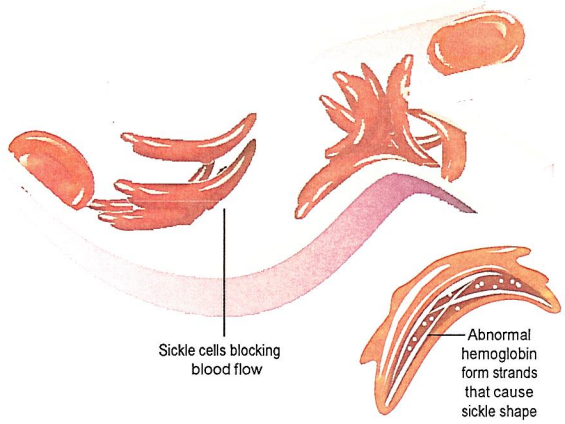


SIGNS AND SYMPTOMS

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.

ABNORMAL, SICKLED RED BLOOD CELLS (SICKLE CELLS)



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.

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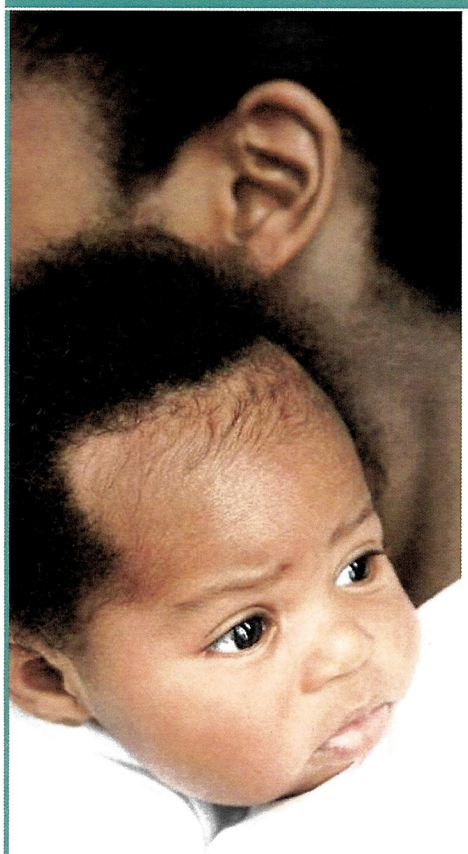
What You Should Know About Sickle Cell Trait



What Is Sickle Cell Trait?

Sickle cell trait (SCT) is not a disease, but having it means that a person has inherited the sickle cell gene from one of his or her parents. People with SCT usually do not have any of the symptoms of sickle cell disease (SCD) and live a normal life.

What Is Sickle Cell Disease?

SCD is a genetic condition that is present at birth. In SCD, the red blood cells become hard and sticky and look like a C-shaped farm tool called a "sickle." The sickle cells die early, which causes a constant shortage of red blood cells. Also, when they travel through small blood vessels, they get stuck and clog the blood flow. This can cause pain and other serious problems. It is inherited when a child receives two sickle cell genes—one from each parent. A person with SCD can pass the disease or SCT on to his or her children.

How Does Someone Get Sickle Cell Trait?

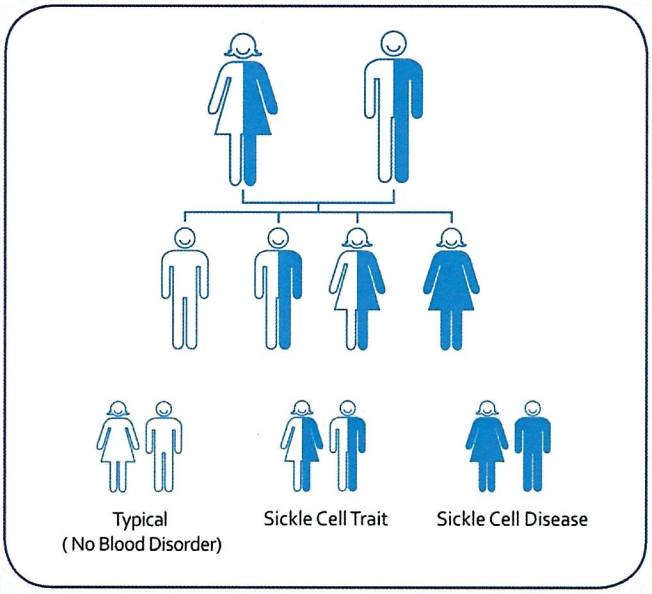
People who have inherited one sickle cell gene and one normal gene have SCT. This means the person won't have the disease, but will be a trait "carrier" and can pass it on to his or her children.

Who Is Affected By Sickle Cell Trait?

SCT affects 1 in 12 Blacks or African Americans in the United States.

- SCT is most common among Blacks or African Americans, but can be found among people whose ancestors come from sub-Saharan Africa; the Western Hemisphere (South America, the Caribbean, and Central America); Saudi Arabia; India; and Mediterranean countries such as Turkey, Greece, and Italy.
- Approximately 3 million people living in the United States have SCT and many are unaware of their status.

What Are The Chances That A Baby Will Have Sickle Cell Trait



- If both parents have SCT, there is a 50% (or 1 in 2) chance that the child also will have SCT if the child inherits the sickle cell gene from one of the parents. Such children will not have symptoms of SCD, but they can pass SCT on to their children.
- If both parents have SCT, there is a 25% (or 1 in 4) chance that the child will have SCD.
- There is the same 25% (or 1 in 4) chance that the child will not have SCD or SCT.
- If one parent has SCT, there is a 50% (or 1 in 2) chance that the child will have SCT and an equal 50% chance that the child will not have SCT.

National Center on Birth Defects and Developmental Disorders
Division of Blood Disorders

CS216728

What Health Complications Are Associated With Sickle Cell Trait?

Most people with SCT do not have any symptoms of SCD, although — in rare cases — people with SCT might experience complications of SCD, such as "pain crises" and, in extreme circumstances, sudden death. More research is needed to find out why some people with SCT have complications and others do not.

In their extreme form and in rare cases, the following conditions could be harmful for people with SCT:

- Increased pressure in the atmosphere (e.g., while scuba diving).
- Low oxygen levels in the air (e.g., when mountain climbing, exercising extremely hard in military boot camp, or training for an athletic competition).
- Dehydration (e.g., too little water in the body).
- High altitudes (e.g., flying, mountain climbing, or visiting a city at a high altitude).

How Will A Person Know If He Or She Has Sickle Cell Trait?

A simple blood test can be done to find out if someone has SCT.

- Testing is available at most hospitals or medical centers, from SCD community-based organizations, or at local health departments.
- A small sample of blood is taken from the finger (a "needle prick") and evaluated in a laboratory.
- If the results of the test reveal that someone has SCT, it is important that he or she know what SCT is, how it can affect him or her, and if and how SCD runs in his or her family.

The best way to find out if and how SCD runs in a person's family is for the person to see a genetic counselor. These professionals have experience with genetic blood disorders. The genetic counselor will look at the person's family history and discuss with him or her what is known about SCD in the person's family. It is best for a person with SCD to learn all he or she can about this disease before deciding to have children.

For more information visit: www.cdc.gov/sicklecell