INTRODUCTION

What Is the Living Well With Sickle Cell Disease: Self-Care Toolkit?

A toolkit is a collection of materials that can be used to help you to manage your health and keep track of important information regarding sickle cell disease (SCD). The Living Well With Sickle Cell Disease: Self-Care Toolkit has multiple uses. It is designed to help you and your caregivers with management of your disease, medical care, services, and health providers. The toolkit also will help communication between the many health providers and service providers that are involved with patient care.

Why Should I Use the Living Well With Sickle Cell Disease: Self-Care Toolkit?

Because many doctors are not familiar with SCD, it is very important for you to take an active role in managing your own care. To make important decisions, you need to know about SCD, understand your treatment options, and then make the best possible choices for your health. Using the tools provided in this toolkit will help you to monitor your health care and manage your disease. Putting together a care notebook or binder that you can take with you wherever and whenever (for example doctor's appointments, emergency room visits, vacation, and the workplace) you need it will help you organize all of your SCD-related medical information in one place so that you can keep track of information over time.

You might want to include the following:

- Doctor contact information.
- Medical appointments.
- Changes in medications or treatments.
- Test results.
- Vaccination and immunization (shots) records.
- Community resources.
- Any other information about your condition (facts found on the Internet, in brochures, and from any other sources of information and support).

By organizing all of your SCD-related information in one place, you can:

- Actively take part in, and advocate for, your own care.
- Work together with the doctors on your medical team.
- Remember new and complex information that is hard to process (when the doctor first tells you about a condition, if the condition worsens, or when treatment changes).
- If you are a teenager or young adult with SCD, you can begin to take responsibility for your own health history and information.

Section 1:
Sickle Cell Disease 101

What Is Sickle Cell Disease?

Sickle cell disease (SCD) is a group of inherited red blood cell disorders.

- Healthy red blood cells are round and they move through small blood vessels carrying oxygen to all parts of the body.
- For someone with SCD, the red blood cells become hard and sticky and look like a C-shaped farm tool called a "sickle".
- Sickle cells die early in comparison to non- sickle cells, which causes a constant shortage of red blood cells.
- Sickle cells can get stuck in small blood vessels and block the flow of blood and oxygen to organs in the body. These changes in cells can cause repeated episodes of severe pain, organ damage, serious infections, or even stroke.

What Causes Sickle Cell Disease?

SCD is inherited in the same way that people get the color of their eyes, skin, and hair.

- A person with SCD is born with it.
- People cannot "catch" SCD from being around a person who has it.

Who Is Affected by Sickle Cell Disease?

- It is estimated that SCD affects 90,000 to 100,000 people in the United States, mainly Blacks or African Americans.
- The disease occurs among about 1 of every 500 Black or African-American births and among about 1 of every 36,000 Hispanic-American births.
- SCD affects millions of people throughout the world and is particularly common among those whose ancestors come from sub-Saharan Africa; regions in the Western Hemisphere (South America, the Caribbean, and Central America); Saudi Arabia; India; and Mediterranean countries such as Turkey, Greece, and Italy.

What Health Problems Does Sickle Cell Disease Cause?

The following are some of the most common complications of SCD:

Pain Episodes or Crises—Sickle cells don't move easily through small blood vessels and can get stuck and clog blood flow. This causes pain that can start suddenly, be mild to severe, and last for any length of time.

Infection—People with SCD, especially infants and children, are more likely to experience harmful infections such as influenza, meningitis (infection of the brain or spinal cord), and hepatitis (infection of the liver).

Hand–Foot Syndrome—Swelling in the hands and feet, often along with a fever, is caused by the sickle cells getting stuck in the blood vessels and blocking the blood from flowing freely through the hands and feet.

Eye Disease—SCD can affect the blood vessels in the eye and lead to long-term damage.

Acute Chest Syndrome—Blockage of the flow of blood to the lungs can cause acute chest syndrome (ACS). ACS is similar to pneumonia; symptoms include but are not limited to chest pain, coughing, difficulty breathing, and fever. It can be life threatening and should be treated in a hospital.

Stroke—Sickle cells can clog blood flow to the brain and cause a stroke. A stroke can result in lifelong disabilities and learning problems.

How Is Sickle Cell Disease Treated?

The goals of treating SCD are to relieve pain and to prevent infections, eye damage, and strokes. There is no single best treatment for all people with SCD. Treatment options are different for each person depending on the symptoms. Treatments can include receiving blood transfusions, receiving intravenous therapy (fluids given into a vein), and medications to help with pain.

- For severe SCD, a medicine called hydroxyurea might be recommended. Research suggests that hydroxyurea can reduce the number of painful episodes and the recurrence of ACS. It also can reduce hospital stays and the need for blood transfusions among adults who have SCD.

Is There a Cure for Sickle Cell Disease?

To date, the only cure for SCD is a bone marrow or stem cell transplant.

- A bone marrow or stem cell transplant is a procedure that takes healthy stem cells from a donor and puts them into someone whose bone marrow is not working properly. These healthy stem cells cause the bone marrow to make new, healthy cells.
- Bone marrow or stem cell transplants are very risky, and can have serious side effects, including death. For the transplant to work, the bone marrow must be a close match.

Section 2:
Living Well With Sickle Cell Disease

Six Steps to Living Well With Sickle Cell Disease

You can live a full life and enjoy most of the activities that other people do. The following tips will help you stay as healthy as possible:

Find good medical care—Sickle cell disease is a complex disease. Good quality medical care from doctors and nurses who know a lot about the disease can help prevent some serious problems. Often, the best choice is a hematologist (a doctor who specializes in blood diseases) working with a team of specialists.

Get regular checkups—Regular health checkups with a primary care doctor can help prevent some serious problems.

Prevent infections—Common illnesses, like influenza, quickly can become dangerous for a person with SCD. The best defense is to take simple steps like washing your hands frequently to help prevent infections. See "Five Tips to Help Prevent Infection" for more information.

Learn healthy habits—Drinking 8 to 10 glasses of water every day and eating healthy food will help to maintain hydration and proper nutrition. People with SCD should maintain a balanced body temperature, getting neither too hot nor too cold. Participating in physical activity to help stay healthy is very important. However, it's essential that you don't overdo it, rest when tired, and drink plenty of water.

Look for clinical studies—New clinical research studies occur frequently and these studies might give you access to new medicines and treatment options.

Get support—Find a patient support group or community-based organization that can provide information, assistance, and support.