Introduction

In 1910, Chicago physician James E. Herrick published a description of oddly shaped blood cells taken from dental student Walter Clement Noel, providing the first detailed in Western medical literature of what has come to be known as sickle cell disease. One hundred years later we know that the sickle-shaped cells are due to a defect in hemoglobin, the protein in red blood cells that carries oxygen throughout the body. A genetic alteration in the hemoglobin molecule causes the body to produce misshapen red blood cells, many of which take the characteristic “C” shape that is the hallmark of sickle cell disease. Sickle cell disease, also known as sickle cell anemia, is inherited. People who have the disease inherit two copies of the sickle gene—one from each parent. The gene codes for the production of an altered form of the protein hemoglobin. Those persons who inherit one copy of the sickle gene, however, do not have sickle cell disease. People who have sickle cell disease do not have the disease, but they carry one of the genes that cause it. Similar to people who have sickle cell disease, people with sickle cell trait can pass the gene to their children. In the United States, sickle cell disease affects an estimated 70,000 to 100,000 people, the majority of whom are African Americans. All states screen newborns for sickle cell disease. In 2010, about 2 million people in the United States have sickle cell trait.

The symptoms and complications of sickle cell disease vary widely. Some people have mild symptoms while others have very severe symptoms and are hospitalized frequently for treatment. Normal red cells pass smoothly through the blood vessels, but sickled cells are stiff and sticky. Sickled cells tend to form clumps that can block blood flow and lead to episodes of extreme pain, known as crises, as well as chronic damage to vital organs. Persons with sickle cell disease have lifelong anemia because their red blood cells survive only about one-tenth as long as cells with normal hemoglobin.

Bone marrow transplants offer a cure to patients who have sickle cell disease. Because of their contributions, we have gained an understanding of the molecular causes of the disease, developed effective approaches for preventing and treating its complications including infection, stroke, and lung and heart failure, and found a small number of people using bone marrow transplants.

The National Heart, Lung, and Blood Institute

The National Heart, Lung, and Blood Institute (NHLBI), which is part of the National Institutes of Health, was founded in 1961 when the NHLBI was established as the National Heart Institute. Since 1972, when the National Sickle Cell Disease Program began, the NHLBI has spent more than $1 billion on sickle cell research. In 2006, the NHLBI began the NHLBI Sickle Cell Disease Program, an initiative intended to address the unmet needs of individuals with sickle cell disease.

Over the past 100 years of sickle cell research we have come to understand that sickle cell disease is a serious public health issue. The NHLBI continues to look ahead to find new and better treatments. Its revitalized research portfolio of basic, clinical, and translational research addresses the genetic factors affecting disease manifestations, regulation of hemoglobin synthesis, development of drugs to increase fetal hemoglobin production, and the development of animal models for preclinical studies. The institute supports research on translation of blood-forming stem cell, gene therapy, a better understanding of and new treatments for pain, optimal uses of blood transfusions, and management of iron overload related to blood transfusions.

The Institute is also leading an effort to develop evidence-based clinical practice guidelines for the care of people who have sickle cell disease, which are expected to be released in 2011. The NHLBI is committed to working with other agencies within the Department of Health and Human Services to disseminate the clinical guidelines with an emphasis on use by primary care practitioners. To ensure that the new guidelines reach their intended audiences, the NHLBI will launch a public awareness and education campaign to focus nationwide attention on sickle cell disease as a serious public health issue.

The NHLBI recognizes that actively engaging patients, families, practitioners, and communities is essential to improving the lives of persons affected by sickle cell disease, and will continue to work with them, community-based groups and scientific organizations to do so.

Sickle Cell Disease and Clinical Trials

The NHLBI sponsors a number of important clinical trials designed to advance the search for better treatments of sickle cell disease. These studies would not be possible without the participation of volunteers who help researchers determine which treatments work. For information on current clinical trials, please visit: http://www.clinicaltrials.gov/.


NHLBI Health Information Center

P.O. Box 20105
Bethesda, MD 20802-0105
Phone: 301-592-8573
TTY: 202-471-9515
Fax: 301-592-8563

Discrimination Prohibited. Under provisions of applicable public laws enacted by Congress since 1964, no person in the United States shall, on the grounds of race, color, national origin, handicap, sex, age, or any other basis included in the relevant adverse action, be excluded from participation in, denied the benefits of, or subjected to discrimination under any program or activity. See the base of each vol. for information on the relevant legal documents. Executive Order 12254 (38 FR 51402) states that federal and federally-funded contractors may not discriminate against any employee or applicant for employment because of race, color, religion, sex, age, or national origin. Therefore the Heart, Lung, and Blood Institute must comply with its own equal opportunity policy and the Executive Order.

Photo credit for 1910 Herrick paper: Archives of Internal Medicine, 1910, VI(5):517-521, Copyright © 1910 American Medical Association. All right reserved.

A Century of Progress: Milestones in Sickle Cell Disease Research

U.S. Department of Health and Human Services
National Institutes of Health

National Heart, Lung, and Blood Institute

Publication No: 10-9767
September 2010

What is Sickle Cell Disease?

Sickle cell anemia, also known as sickle cell disease, is inherited. People who have the disease inherit two copies of the sickle cell gene—one from each parent. The gene codes for production of an abnormal form of the protein hemoglobin, and as a result of this genetic abnormality persons with sickle cell disease have a reduced ability to transport oxygen and nutrients to vital organs. Persons with sickle cell disease have life-long extreme pain, known as crises, as well as chronic damage to vital organs. Persons with sickle cell disease have life-long anemia because their red blood cells survive only about one-tenth as long as cells with normal hemoglobin.

In 1910, Chicago physician James E. Herrick published a description of oddly shaped blood cells taken from dental student Walter Clement Noel, providing the first detailed in Western medical literature of what has come to be known as sickle cell disease. One hundred years later we know that the sickle-shaped cells are due to a defect in hemoglobin, the protein in red blood cells that carries oxygen throughout the body. A genetic alteration in the hemoglobin molecule causes the body to produce misshapen red blood cells, many of which take the characteristic “C” shape that is the hallmark of sickle cell disease.

The NHLBI recognizes that actively engaging patients, families, practitioners, and communities is essential to improving the lives of persons affected by sickle cell disease, and will continue to work with them, community-based groups and scientific organizations to do so.

Sickle Cell Disease and Clinical Trials

The NHLBI sponsors a number of important clinical trials designed to advance the search for better treatments of sickle cell disease. These studies would not be possible without the participation of volunteers who help researchers determine which treatments work. For information on current clinical trials, please visit: http://www.clinicaltrials.gov/.


NHLBI Health Information Center

P.O. Box 20105
Bethesda, MD 20802-0105
Phone: 301-592-8573
TTY: 202-471-9515
Fax: 301-592-8563

Discrimination Prohibited. Under provisions of applicable public laws enacted by Congress since 1964, no person in the United States shall, on the grounds of race, color, national origin, handicap, sex, age, or any other basis included in the relevant adverse action, be excluded from participation in, denied the benefits of, or subjected to discrimination under any program or activity. See the base of each vol. for information on the relevant legal documents. Executive Order 12254 (38 FR 51402) states that federal and federally-funded contractors may not discriminate against any employee or applicant for employment because of race, color, religion, sex, age, or national origin. Therefore the Heart, Lung, and Blood Institute must comply with its own equal opportunity policy and the Executive Order.

Photo credit for 1910 Herrick paper: Archives of Internal Medicine, 1910, VI(5):517-521, Copyright © 1910 American Medical Association. All right reserved.

A Century of Progress: Milestones in Sickle Cell Disease Research

U.S. Department of Health and Human Services
National Institutes of Health

National Heart, Lung, and Blood Institute

Publication No: 10-9767
September 2010

What is Sickle Cell Disease?

Sickle cell anemia, also known as sickle cell disease, is inherited. People who have the disease inherit two copies of the sickle cell gene—one from each parent. The gene codes for production of an abnormal form of the protein hemoglobin, and as a result of this genetic abnormality persons with sickle cell disease have a reduced ability to transport oxygen and nutrients to vital organs. Persons with sickle cell disease have life-long extreme pain, known as crises, as well as chronic damage to vital organs. Persons with sickle cell disease have life-long anemia because their red blood cells survive only about one-tenth as long as cells with normal hemoglobin.

In 1910, Chicago physician James E. Herrick published a description of oddly shaped blood cells taken from dental student Walter Clement Noel, providing the first detailed in Western medical literature of what has come to be known as sickle cell disease. One hundred years later we know that the sickle-shaped cells are due to a defect in hemoglobin, the protein in red blood cells that carries oxygen throughout the body. A genetic alteration in the hemoglobin molecule causes the body to produce misshapen red blood cells, many of which take the characteristic “C” shape that is the hallmark of sickle cell disease.

The NHLBI recognizes that actively engaging patients, families, practitioners, and communities is essential to improving the lives of persons affected by sickle cell disease, and will continue to work with them, community-based groups and scientific organizations to do so.

Sickle Cell Disease and Clinical Trials

The NHLBI sponsors a number of important clinical trials designed to advance the search for better treatments of sickle cell disease. These studies would not be possible without the participation of volunteers who help researchers determine which treatments work. For information on current clinical trials, please visit: http://www.clinicaltrials.gov/.


NHLBI Health Information Center

P.O. Box 20105
Bethesda, MD 20802-0105
Phone: 301-592-8573
TTY: 202-471-9515
Fax: 301-592-8563

Discrimination Prohibited. Under provisions of applicable public laws enacted by Congress since 1964, no person in the United States shall, on the grounds of race, color, national origin, handicap, sex, age, or any other basis included in the relevant adverse action, be excluded from participation in, denied the benefits of, or subjected to discrimination under any program or activity. See the base of each vol. for information on the relevant legal documents. Executive Order 12254 (38 FR 51402) states that federal and federally-funded contractors may not discriminate against any employee or applicant for employment because of race, color, religion, sex, age, or national origin. Therefore the Heart, Lung, and Blood Institute must comply with its own equal opportunity policy and the Executive Order.

Photo credit for 1910 Herrick paper: Archives of Internal Medicine, 1910, VI(5):517-521, Copyright © 1910 American Medical Association. All right reserved.
**1940-1945**

Sickle cell disease and its clinical implications were widely discussed. The National Heart Institute established for newborn screening to identify sickle cell disease.

---

**1940**

- Painful sickle cell “crises” suggested by Dr. John Caveness, of the Journal of the National Medical Association.

---

**1945**

- Sickle cell anemia: clinical implications of the disease.

---

**1946**

- The National Heart Institute established first grants include $40,000 to James, to study the clinical course of sickle cell disease.

---

**1947**

- Dr. Robert B. Scott, the first black president of the National Medical Association, discusses sickle cell disease.

---

**1948**

- The National Heart Institute established.

---

**1949**

- The disease of the red blood cells.

---

**1950**

- Sickle cell disease: a cause of major concern.

---

**1951**

- The National Heart Institute established.

---

**1952**

- The first three-dimensional structure of the hemoglobin molecule is determined.

---

**1953**

- Scientists show abnormality of the sickle cell.

---

**1954**

- Scientists show abnormality of the sickle cell.

---

**1955**

- Scientists show abnormality of the sickle cell.

---

**1956**

- Scientists show abnormality of the sickle cell.

---

**1957**

- Scientists show abnormality of the sickle cell.

---

**1958**

- Scientists show abnormality of the sickle cell.

---

**1959**

- Scientists show abnormality of the sickle cell.

---

**1960**

- Scientists show abnormality of the sickle cell.

---

**1961**

- Scientists show abnormality of the sickle cell.

---

**1962**

- Scientists show abnormality of the sickle cell.

---

**1963**

- Scientists show abnormality of the sickle cell.

---

**1964**

- Scientists show abnormality of the sickle cell.

---

**1965**

- Scientists show abnormality of the sickle cell.

---

**1966**

- Scientists show abnormality of the sickle cell.

---

**1967**

- Scientists show abnormality of the sickle cell.

---

**1968**

- Scientists show abnormality of the sickle cell.

---

**1969**

- Scientists show abnormality of the sickle cell.

---

**1970**

- Scientists show abnormality of the sickle cell.

---

**1971**

- Scientists show abnormality of the sickle cell.

---

**1972**

- Scientists show abnormality of the sickle cell.

---

**1973**

- Scientists show abnormality of the sickle cell.

---

**1974**

- Scientists show abnormality of the sickle cell.

---

**1975**

- Scientists show abnormality of the sickle cell.

---

**1976**

- Scientists show abnormality of the sickle cell.

---

**1977**

- Scientists show abnormality of the sickle cell.

---

**1978**

- Scientists show abnormality of the sickle cell.

---

**1979**

- Scientists show abnormality of the sickle cell.

---

**1980**

- Scientists show abnormality of the sickle cell.

---

**1981**

- Scientists show abnormality of the sickle cell.

---

**1982**

- Scientists show abnormality of the sickle cell.

---

**1983**

- Scientists show abnormality of the sickle cell.

---

**1984**

- Scientists show abnormality of the sickle cell.

---

**1985**

- Scientists show abnormality of the sickle cell.

---

**1986**

- Scientists show abnormality of the sickle cell.

---

**1987**

- Scientists show abnormality of the sickle cell.

---

**1988**

- Scientists show abnormality of the sickle cell.

---

**1989**

- Scientists show abnormality of the sickle cell.

---

**1990**

- Scientists show abnormality of the sickle cell.

---

**1991**

- Scientists show abnormality of the sickle cell.

---

**1992**

- Scientists show abnormality of the sickle cell.

---

**1993**

- Scientists show abnormality of the sickle cell.

---

**1994**

- Scientists show abnormality of the sickle cell.

---

**1995**

- Scientists show abnormality of the sickle cell.

---

**1996**

- Scientists show abnormality of the sickle cell.

---

**1997**

- Scientists show abnormality of the sickle cell.

---

**1998**

- Scientists show abnormality of the sickle cell.

---

**1999**

- Scientists show abnormality of the sickle cell.

---

**2000**

- Scientists show abnormality of the sickle cell.

---

**2001**

- Scientists show abnormality of the sickle cell.

---

**2002**

- Scientists show abnormality of the sickle cell.

---

**2003**

- Scientists show abnormality of the sickle cell.

---

**2004**

- Scientists show abnormality of the sickle cell.

---

**2005**

- Scientists show abnormality of the sickle cell.

---

**2006**

- Scientists show abnormality of the sickle cell.

---

**2007**

- Scientists show abnormality of the sickle cell.

---

**2008**

- Scientists show abnormality of the sickle cell.

---

**2009**

- Scientists show abnormality of the sickle cell.

---

**2010**

- Scientists show abnormality of the sickle cell.

---

**2011**

- Scientists show abnormality of the sickle cell.

---

**2012**

- Scientists show abnormality of the sickle cell.

---

**2013**

- Scientists show abnormality of the sickle cell.

---

**2014**

- Scientists show abnormality of the sickle cell.

---

**2015**

- Scientists show abnormality of the sickle cell.

---

**2016**

- Scientists show abnormality of the sickle cell.

---

**2017**

- Scientists show abnormality of the sickle cell.

---

**2018**

- Scientists show abnormality of the sickle cell.

---

**2019**

- Scientists show abnormality of the sickle cell.

---

**2020**

- Scientists show abnormality of the sickle cell.

---

**2021**

- Scientists show abnormality of the sickle cell.

---

**2022**

- Scientists show abnormality of the sickle cell.

---

**2023**

- Scientists show abnormality of the sickle cell.

---

**2024**

- Scientists show abnormality of the sickle cell.

---

**2025**

- Scientists show abnormality of the sickle cell.

---