



Sickle Cell Disease

Sickle cell disease is one of a group of inherited red blood cell disorders called hemoglobinopathies. In sickle cell disease, 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 such as infection, acute chest syndrome and stroke. It is estimated that sickle cell disease affects 90,000 to 100,000 Americans.

2012 Accomplishments

- Developed the Registry and Surveillance for Hemoglobinopathies (RuSH) "Strategies from the Field: Data Collection" and "Strategies from the Field: Health Promotion." The publications, which are available on CDC's website, showcase the various methods that can be adapted by states to develop state-specific population-based data collection and community engagement to improve the lives of people with sickle cell disease and thalassemia.
- Implemented Public Health Research Epidemiology and Surveillance for Hemoglobinopathies (PHRESH), a demonstration project that focuses on three areas: surveillance, health promotion and prevention awareness, and laboratory capacity building. The project is intended to expand upon, and learn more about, the data collected from RuSH and is being piloted in California, Mississippi, and Georgia.
- Developed and submitted a manuscript entitled, "*Invasive Pneumococcal Disease among Children with and without Sickle Cell Disease in the United States, 1998-2009*" to the Pediatric Infectious Disease Journal. The manuscript describes an assessment of trends in invasive pneumococcal disease in children less than 18 years of age with sickle cell disease.

AJ's Story



At 19 years old, AJ is an accomplished singer and sickle cell disease advocate, and doesn't let his condition keep him from reaching his goals.

To read AJ's story, visit:
www.cdc.gov/ncbddd/SickleCell/stories.htm
1

2012 Accomplishments (continued)

- Hosted a monthly Public Health Webinar Series on Hemoglobinopathies. The purpose of this webinar series is to offer a hemoglobinopathies learning collaborative platform for providers, general public, educators, and scientists.
- Partnered with the Sickle Cell Disease Association of America and the National Baptist Congress of Christian Education to develop and disseminate a new video entitled, “Sickle Cell Disease Education and Awareness in Your Community.” The video raises awareness and promotes education of sickle cell disease by illustrating the public health role, community involvement, and individual level impact of sickle cell disease in the United States.
- Hosted the 2nd National Conference on Blood Disorders in Public Health. The purpose of this conference was to promote the health and improve outcomes among people with blood disorders.

Looking to the Future

There is still much to learn about sickle cell disease and how to prevent and manage complications associated with the disease. The impact of genetic, environmental, behavioral, and other risk factors on sickle cell disease is not fully understood. An estimated 90% of people with sickle cell disease are unable to attain the resources they need to improve or maintain their health and proven therapies are not being used. It is estimated that 30% or less of eligible people with sickle cell disease are treated with hydroxyurea, which is known to reduce pain crises, stroke, pulmonary complications and disability.

Strengthening CDC’s sickle cell disease programs and activities in the areas of surveillance, health promotion, technical assistance to community-based organizations and state health departments can fill the public health needs essential for saving, lengthening, and improving the quality of lives of people with sickle cell disease.

Collaborating with our many public health partners – other federal and state agencies, academia, and professional and community-based organizations – The National Center on Birth Defects and Developmental Disabilities’ Division of Blood Disorders improves the lives of people at risk or affected by sickle cell disease.

Did You Know?

- In the U.S., 2,000 infants are born with sickle cell disease each year and an estimated 90,000 to 100,000 Americans are living with sickle cell disease.
- 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.
- During 2005, medical expenditures for children with sickle cell disease averaged \$11,702 for children with Medicaid coverage and \$14,772 for children with employer-sponsored insurance.

Notable Scientific Publications

Wang Y, Kennedy J, Caggana M, et al. Sickle cell disease incidence among newborns in New York State by maternal race/ethnicity and nativity. *Genet Med*. 2012 Sep 27.

Grosse SD, Atrash HK, Odame I, et al. The Jamaican historical experience of the impact of educational interventions on sickle cell disease child mortality. *Am J Prev Med*. 2012 Jun;42(6):e101-3.

Bean CJ, Boulet SL, Ellingsen D, et al. Heme oxygenase-1 gene promoter polymorphism is associated with reduced incidence of acute chest syndrome among children with sickle cell disease. *Blood*. 2012 Nov 1;120(18):3822-8.

Thangarajh M, Yang G, Fuchs D, et al. Magnetic resonance angiography-defined intracranial vasculopathy is associated with silent cerebral infarcts and glucose-6-phosphate dehydrogenase mutation in children with sickle cell anaemia. *Br J Haematol*. 2012 Nov;159(3):352-9.

Bae HT, Baldwin CT, Sebastiani P, et al. Meta-analysis of 2040 sickle cell anemia patients: BCL11A and HBS1L-MYB are the major modifiers of HbF in African Americans. *Blood*. 2012 Aug 30;120(9):1961-2.

Milton JN, Sebastiani P, Solovieff N, et al. A genome-wide association study of total bilirubin and cholelithiasis risk in sickle cell anemia. *PLoS One*. 2012;7(4):e34741.

McClellan, A. C., Luthi, J.-C., Lynch, J. R., et al. (2012), High one year mortality in adults with sickle cell disease and end-stage renal disease. *British Journal of Haematology*. doi: 10.1111/bjh.12024.

To view the annual report online, visit:
www.cdc.gov/ncbddd/2012AnnualReport

For more information about sickle cell disease, visit:
www.cdc.gov/SickleCell