

Sickle Cell Disease in Georgia

FINDINGS FROM RuSH FACTS FOR PROVIDERS

What is RuSH?

- Registry and Surveillance System for Hemoglobinopathies (RuSH) was a pilot project of the Centers for Disease Control and Prevention (CDC), with the National Institutes of Health's National Heart, Lung, and Blood Institute.
- The overall purpose of RuSH was to collect state-specific, population-based data on people with sickle cell disease (SCD) and thalassemia in order to provide accurate updated information to the public.
- Seven states were funded to participate in data collection: California, Florida, Georgia, Michigan, New York, North Carolina, and Pennsylvania.

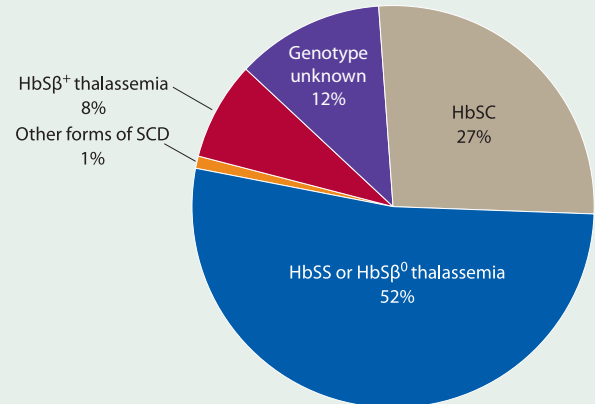
What is Sickle Cell Disease?

- SCD is a group of inherited conditions that affect hemoglobin, a protein that allows red blood cells to carry oxygen to all parts of the body.
- People with ancestry from parts of Africa, Asia and the Mediterranean are among the most likely to be born with these conditions.
- Healthy red blood cells are round, and they move through small blood vessels to carry oxygen to all parts of the body. In SCD, the red blood cells become hard and sticky and look like a C-shaped farm tool called a "sickle."
- These cells can get stuck in the blood vessels and block the normal flow of oxygen throughout the body. This leads to a variety of health problems.
- The most common types of SCD are:
 - *Hemoglobin SS Disease (HbSS)*: People who have this form of SCD inherit two sickle cell hemoglobin genes ("S"), one from each parent. This is commonly called *sickle cell anemia* and is usually the most severe form of the disease.
 - *Hemoglobin SC Disease (HbSC)*: People who have this form of SCD inherit a sickle cell hemoglobin gene ("S") from one parent and from the other parent a gene for abnormal hemoglobin called "C". This is usually a milder form of SCD.
 - *Hemoglobin S beta thalassemia (HbS beta thalassemia)*: People who have this form of SCD inherit one sickle cell hemoglobin gene ("S") from one parent and one gene for beta thalassemia, another type of anemia, from the other parent. There are two types of beta thalassemia: "0" and "+". Those with HbS beta⁰-thalassemia usually have a more severe form of SCD. People with HbS beta⁺-thalassemia tend to have a milder form of SCD.

The Sickle Cell Population in Georgia

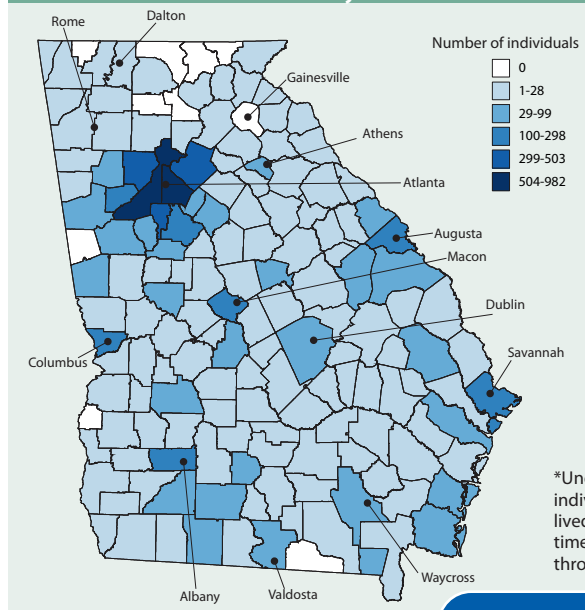
SCD occurs in individuals of all geographic and ethnic backgrounds. We estimate that 97% or more of Georgia newborns with SCD are Black or African-American, and roughly 2% overall are of Hispanic-American ethnicity. This means that about 1 out of every 295 Black or African-American babies born in Georgia from 2004 through 2008 had some form of SCD.

Types of SCD in babies born in Georgia, 2004-2008



Through our surveillance efforts, we found 7,299 individuals with confirmed or probable SCD in Georgia. Of these, 4,288 had a diagnosis confirmed by a clinical center or through newborn screening. The rest were identified because they had a sickle cell diagnosis in one of the administrative datasets (hospital or public insurance claims data) as well as health care utilization consistent with SCD.¹ These 7,299 individuals live in virtually every county throughout the state of Georgia.

Number of individuals with SCD in Georgia counties identified by surveillance*

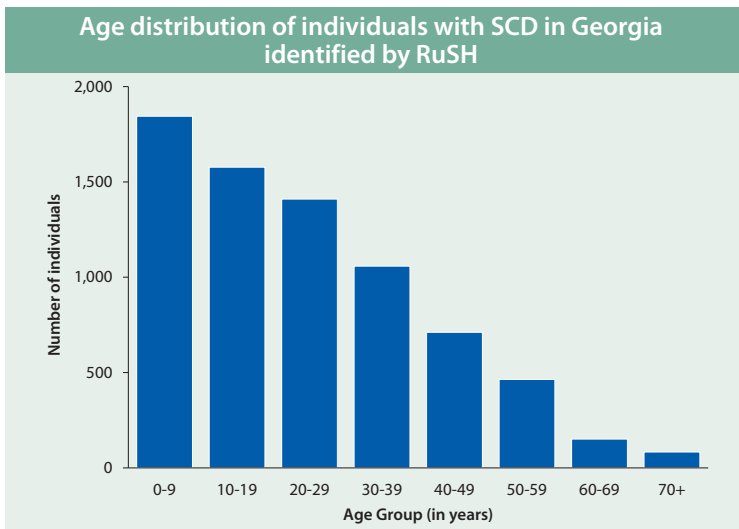


¹Based on CDC RuSH definition

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



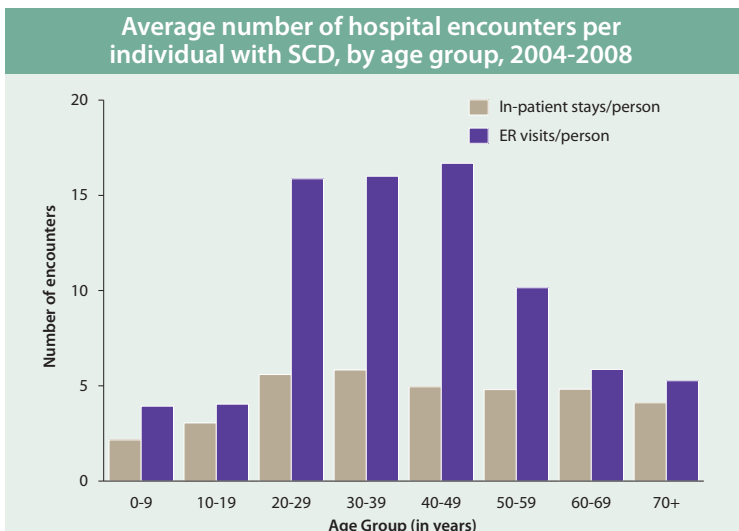
We identified individuals living with SCD from newborns to those over 70 years of age; the figure below shows the age distribution of identified cases. Because our surveillance methods relied heavily on utilization data from newborn screening follow-up facilities and public health insurance programs, we were more likely to identify children living with the disease than their adult counterparts. The decreased prevalence by age of people living with SCD illustrated by the figure is in part due to our surveillance methods and not only a reflection of early mortality from the disease.



Health and Health Care

Again, because our surveillance methods relied on health care utilization data and insurance claims, individuals with SCD who did not seek care—some who were uninsured and some whose disease was well managed—were less likely captured in our results. The following health and health care utilization results, therefore, likely represent those with greater health needs.

Of the 7,299 individuals with SCD identified, we have access to utilization data for 94 percent (6,881) of them. Twenty-six percent (1,798/6,881) had no hospitalizations, and 16 percent (1,131/6,881) had no ER visits during the five-year period from 2004 to 2008. The figure below illustrates the five-year average number of hospital admissions and ER visits by age group. This health care utilization



shows a pronounced increase after childhood. While children age 0-19 averaged roughly four ER visits and three hospitalizations over five years, those age 20 through 49 had more than fifteen ER visits in addition to a greater frequency of hospital admissions. Although not assessed by our surveillance, this may be in part related to the natural history of the condition; factors directly related to the transition from parental care to independence, from pediatric to adult medical care, and to changes in health insurance coverage. Transition to independent self-care from parent directed management may require some learning. In addition, a break in the primary care medical home may cause discontinuity in crucial primary prevention and clinical support for patients with SCD.

What you can do

- Learn more about SCD:
 - Sickle Cell Information Center website: scinfo.org
 - Comprehensive Sickle Cell Center at Georgia Regents University: 706-721-2171
 - Georgia Comprehensive Sickle Cell Center at Grady Health System: 404-616-3572
 - Aflac Cancer and Blood Disorders Center of Children's Healthcare of Atlanta: 404-785-1200
- Report babies and children with SCD to the Department of Public Health (sickle cell is a notifiable disease for children through age 18 in Georgia): health.state.ga.us/epi/disease/report; 404-657-2588
- Refer SCD patients and families for information and support services: Sickle Cell Foundation of Georgia, Inc.: www.sicklecclga.org; 404-755-1641; 800-326-5287 (toll-free)
- Educate patients on the role of newborn screening: Georgia Department of Public Health Newborn Screening Program health.state.ga.us/programs/nsmscd; 404-657-4143
- Submit samples for confirmatory testing to Georgia's reference laboratory for hemoglobinopathies, the Titus HJ Huisman Hemoglobinopathy Laboratory at Georgia Regents University: georgiahealth.edu/centers/sickleccl; 706-721-9640
- Learn more about the RuSH Project: Angela Snyder, angiesnyder@gsu.edu