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Using surveillance to determine the number of individuals with sickle cell disease in California and Georgia, 2005–2016

Aika Aluc[a,b], **Mei Zhou**[c], **Susan T. Paulukonis**[d], **Angela B. Snyder**[c,e], **David Wong**[a], **Mary M. Hulihan**[f]

[a]Office of Minority Health, U.S. Department of Health & Human Services, Rockville, Maryland, USA;

[b]Oak Ridge Institute for Science and Education, Tennessee, USA;

[c]Andrew Young School of Policy Studies, Georgia Health Policy Center, Georgia State University, Atlanta, USA;

[d]Tracking California Program, Public Health Institute, Richmond, Virginia, USA;

[e]Andrew Young School of Policy Studies, Department of Public Management and Policy, Georgia State University, Atlanta, USA;

[f]Division of Blood Disorders, National Center for Birth Defects and Disabilities, CDC, Atlanta, Georgia, USA

Introduction

Sickle cell disease (SCD) is an inherited blood disorder that affects the shape and function of red blood cells. It is most common among people of African descent and affects one of every 365 African American newborns and approximately 100,000 persons in the United States.[1] Major complications include debilitating pain, infection, stroke, and organ damage. A national surveillance system for SCD does not exist; however, two states, California and Georgia, conduct population-level SCD surveillance. These systems comprise the CDC Sickle Cell Data Collection (SCDC) program, which includes comprehensive data linkages from multiple data systems.[2] These data, which include information about demographics, payer, and health care utilization, identify opportunities for improving access to care. They may be used to make informed decisions about locations for new clinics staffed by providers who are knowledgeable about SCD, its complications, and available treatments. This report presents findings from SCDC on the birth prevalence and number of individuals with SCD in California and Georgia from 2005 to 2016.

CONTACT Mary M. Hulihan, ibx5@cdc.gov, Division of Blood Disorders, National Center for Birth Defects and Disabilities, CDC, 4770 Buford Hwy, Atlanta, GA 30341, USA.

Disclosure statement

No potential conflict of interest was reported by the authors.

Materials and methods

The SCDC programs in both California and Georgia used a comparable methodology. The programs ascertained individual-level health and mortality data on individuals with SCD through linkage of multiple population-based data sources: newborn screening, emergency department (ED) discharge, hospital discharge, state Medicaid, and death records, and also linkage with medical records data from SCD clinical centers.[2] Individuals in the various data sources were identified as having SCD using a conservative and validated (in individuals <21-year old) case definition that required either a laboratory-confirmed diagnosis of SCD or the occurrence of three of more healthcare encounters with an SCD-specific ICD-9-CM or ICD-10-CM diagnosis code over a 5-year period.[3] This analysis covered individuals residing in each state at any time during 2005 through 2016. SCDC data were analyzed to determine the total and annual numbers of infants with SCD born in each state. Data were also assessed according to demographic factors, SCD genotype, and mother's county of residence at time of birth. For this analysis, information from CDC WONDER[4] was used for the calculation of SCD birth rate per 10,000 live births and 10,000 Black births across three four-year periods, for those counties with 40 or more SCD births during 2005–2016. SCDC data were also used to count the number of children and adults with SCD identified in each of the states, by sex and age, per year.

Results

During 2005 through 2016, the average annual number of newborns with SCD born to state residents was 90 (range 68–117; total 1,075) in California and 157 (range 134–223; total 1,880) in Georgia. Forty-eight percent of the newborns were female. Approximately 12% of babies identified with SCD in California were Hispanic; information about ethnicity was not available in Georgia. Fifty-six percent had sickle cell anemia (Hemoglobin (Hb) S/S or S/β^0-thalassemia; California: 55%, Georgia: 57%); 28% had Hb S/C (California and Georgia: 28%); 8% had Hb S/β^+-thalassemia (California: 10%, Georgia: 7%); 3% had another form of SCD (California: 6%, Georgia: 1%); and the type of SCD was unknown in 4% (California: 0%, Georgia: 7%). There were 1.7 SCD births per 10,000 live births in California and 27.4 per 10,000 live births to Black or African-American mothers during 2005–2016. In Georgia, there were 11.4 SCD births per 10,000 live births and 31.9 per 10,000 live births to Black or African American mothers. Fifty five percent (32/58) of the counties in California and 76% (121/159) of the counties in Georgia had a least one SCD birth during the 12-year period. A single county in California, Los Angeles County, was the site of 39% of all SCD births in the state while two counties in Georgia, Fulton, and DeKalb, accounted for 27% of the SCD births in that state. Although the SCD birth rates remained stable at the state level, there was wide variation at the county level (Table 1). In California, for example, there were increases in the SCD birth rate in Riverside and San Bernardino counties, while both Alameda and San Joaquin counties saw declines. Between 2010 and 2015, an average of 5,022 individuals in California and 7,749 individuals in Georgia were identified with SCD each year (Figure 1). In California, 37% of these individuals were <20-year old, 51% were 20- to 49-year old, and 12% were 50 years and older. In Georgia, the percentages were 46%, 45%, and 9% for these age groups. Fifty-six percent of the individuals with SCD identified in both states were female.

Discussion

The SCDC surveillance data allow for a unique understanding about the number of individuals with SCD in California and Georgia, as compared to estimates last published in 2010.[1,5] Brousseau et al estimated 6,474 individuals with SCD in California and 5,890 in Georgia. Dr. Hassell's estimates ranged from 4,240 to 4,707 for California and 4,981 to 5,797 for Georgia. In general, both of those publications applied the birth prevalence rates for SCD to US Census data and adjusted for early mortality based on age and sickle cell type. The data presented in this report differ in that they are based on counts of individuals, collected in a multisource data system with a validated (for individuals <21-year old) case definition, rather than estimates.

SCDC aims to include all individuals with SCD, regardless of their disease severity, where they live, or where they receive their care. Many individuals with SCD continue to face challenges finding and accessing a knowledgeable physician, especially adults and those living in non-urban areas where a majority of the health care centers with a full array of specialty providers are located. As such, these data may be used to identify opportunities for improving access to care for individuals with SCD, such as locations for new clinics staffed by providers who are knowledgeable about SCD, its complications, and available treatments. Conversely, the data may be used to target providers who are already seeing patients with SCD and could benefit from educational resources about the latest developments in clinical research, guidelines, and trials that may enhance the care they deliver.

The findings in this report are subject to at least two limitations. First, because SCDC relies on administrative data (both Medicaid and hospital/ED claims) to identify individuals with an SCD diagnosis code, individuals without hospital level care, those who are uninsured, or those who are privately insured, as well as individuals without any health care use may be missed; however, by using multiple years of data, the magnitude of undercounting is reduced. Also, at this time, SCDC is able to track individuals with SCD only as long as they stay in California or Georgia; mobility across state borders remains a challenge. Finally, this analysis is limited to only two states. Due to differences in health care systems, health care policy, populations, and resources among states, it is not expected that these results are representative of the entire nation. Establishing the SCDC system in a larger number of states and continuing the project over an extended period of time would allow for a richer and more complete understanding of the similarities and differences across states, in terms of SCD.

The longitudinal data collected by the SCDC program allow for future work to better understand what happens to the individuals identified with SCD as they age and live with a chronic condition. Future opportunities to provide high quality care for all those with the disease as the population transitions to adulthood merit further exploration.

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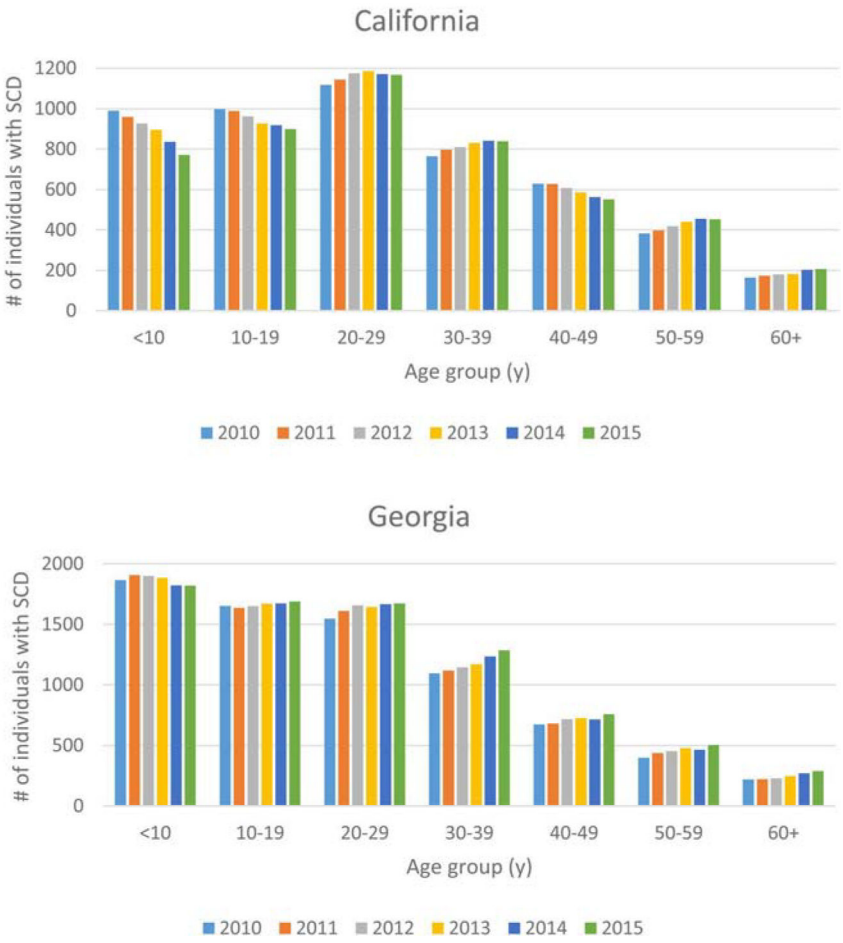

Figure 1.

Number of individuals with sickle cell disease identified in California and Georgia, by age group, 2010–2015.

Table 1.

Sickle cell disease birth rate, by county, California and Georgia, 2005–2016.

	2005–2008			2009–2012			2013–2016		
	SCD births, *n*	**SCD birth rate/10,000 live births**	**SCD birth rate/ 10,000 live Black births**	**SCD births, *n***	**SCD birth rate/10,000 live births**	**SCD birth rate/ 10,000 live Black births**	**SCD births, *n***	**SCD birth rate/10,000 live births**	**SCD birth rate/ 10,000 live Black births**
CALIFORNIA TOTAL	372	1.67	27.51	364	1.78	27.58	339	1.71	27.12
Alameda County, CA	30	3.55	27.57	28	3.58	27.35	21	2.70	23.98
Fresno County, CA	13	1.94	34.19	13	2.01	33.21	14	2.26	37.59
Los Angeles County, CA	160	2.66	33.24	137	2.56	30.19	121	2.39	29.19
Riverside County, CA	14	1.06	18.31	20	1.62	25.16	31	2.56	37.51
Sacramento County, CA	22	2.54	21.11	25	3.12	23.29	20	2.56	19.35
San Bernardino County, CA	27	1.97	21.95	49	3.93	39.60	40	3.25	32.63
San Diego County, CA	24	1.28	22.48	21	1.18	20.36	19	1.09	18.87
San Joaquin County, CA	18	3.92	48.75	13	3.10	35.24	11	2.74	30.43
GEORGIA TOTAL	677	11.50	33.72	616	11.45	31.33	587	11.26	30.58
Bibb County, GA	12	11.43	18.87	19	19.34	30.42	11	11.94	18.65
Chatham County, GA	25	15.62	32.55	40	24.77	54.53	23	14.34	31.90
Clayton County, GA	58	29.82	48.18	33	20.70	30.07	39	26.28	36.36
Cobb County, GA	40	9.04	33.36	51	13.23	41.19	49	12.76	38.78
DeKalb County, GA	97	21.20	38.43	71	16.12	29.26	74	16.87	31.41
Fulton County, GA	86	15.00	30.57	85	15.31	29.10	93	17.34	32.54
Gwinnett County, GA	55	10.26	43.76	48	10.53	35.77	56	12.36	39.58
Muscogee County, GA	22	17.01	33.17	16	11.86	23.35	19	15.45	30.75
Richmond County, GA	16	12.09	20.13	24	19.15	31.70	25	19.88	34.08

*Counties with 40 or more births during 2005–2016.

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