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ABSTRACT

Objective. Population-based surveillance data from California and Georgia for years 2004 through 2008 were linked to state death record files to determine the all-cause death rate among 12,143 patients identified with sickle cell disease (SCD).

Methods. All-cause death rates, by age, among these SCD patients were compared with all-cause death rates among both African Americans and the total population in the two states. All-cause death rates were also compared with death rates for SCD derived from publicly available death records: the compressed mortality files and multiple cause of death files.

Results. Of 12,143 patients identified with SCD, 615 patients died. The all-cause mortality rate for the SCD population was lower than the all-cause mortality rate among African Americans and similar to the total population all-cause mortality rates from birth through age 4 years, but the rate was higher among those with SCD than both the African American and total population rates from ages 5 through 74 years. The count of deceased patients identified by using population-based surveillance data (*n*=615) was more than twice as high as the count identified in compressed mortality files using SCD as the underlying cause of death alone (*n*=297).

Conclusion. Accurate assessment of all-cause mortality and age at death requires long-term surveillance via population-based registries of patients with accurately diagnosed SCD.

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Ongoing public health surveillance of sickle cell disease (SCD) is critical to understanding its course and outcomes. Such surveillance is vital for medical management, clinical and translational research, new therapy development, public health planning, and patient, family, and provider education.

Multiple research studies have used data collected through limited surveillance activities to better understand age at death for patients with SCD. Studies published in the 1970s showed a poor prognosis for patients with SCD, with a median age at death of approximately 14 years, high mortality in the first five years of life, and only about 15% of deaths occurring after age 30 years.[1] Results from a 1978 study based on a newborn screening cohort in Jamaica found that 13% of children with hemoglobin SS disease and 5% of children with hemoglobin SC disease died in the first two years of life.[2] Follow-up studies in Jamaica confirmed this observation and showed that only 25% of deaths among those with SCD occurred in patients older than 30 years of age.[3]

The Cooperative Study of Sickle Cell Disease, which followed SCD patients in the United States from 1978 through 1988, reported patient survival in its academic center cohort, with a median age at death of 48 years for women and 42 years for men with hemoglobin SS disease, and 68 years for women and 60 years for men with hemoglobin SC disease.[4] This study also reported that 50% of hemoglobin SS disease deaths occurred in patients older than 45 years of age. A 2001 study, using excess mortality simulation of a Jamaican cohort, predicted that 50% of SCD deaths would occur in those older than 53 years of age.[5] Additional studies based on newborn screening cohorts enrolled in comprehensive care systems in the United States (published in 2004 and 2010), the United Kingdom (published in 2007), and Brazil (published in 2014) indicated that mortality in the first five years of life was significantly lower than in the 1978 Jamaican report, with the expectation that 86% to 98% of children with SCD would live to adulthood.[2,6–9]

However, not all recent research indicates similar findings in SCD mortality. A study using data from the National Center for Health Statistics' (NCHS's) compressed mortality (CM) files, 1999 through 2006, estimated the mean age at death for patients with SCD to be 39 years in 2006.[10] In that study, among the 483 SCD deaths in the United States reported during that year, 9% occurred in those aged 20 years or younger and 35% occurred in those older than 45 years of age. A study using NCHS's multiple cause of death (MCOD) files, 1979 through 2005, to estimate age at death and death rates similarly found median ages at death among women and men with SCD to be 42 and 38 years, respectively.[11]

One important caveat to previous mortality studies is that researchers relied on death certificate reporting of SCD[10,11] or on populations seen at high-volume hematology clinics.[2–9] Studies based on death certificates assumed accurate and complete coding in the death record, while the clinic-based studies may have been biased to include the sickest SCD patients, who are more likely to be seen at specialty care centers. To address these limitations, we used data on SCD patients from California and Georgia who were identified during the Registry and Surveillance System for Hemoglobinopathies project and linked them to all state death record files to estimate the all-cause mortality rate and mean and median ages at death for SCD in these two states.[12]

METHODS

The National Institutes of Health-funded and Centers for Disease Control and Prevention (CDC)-directed Registry and Surveillance System for Hemoglobinopathies project collected and linked population-based surveillance data from a variety of sources in California and Georgia for 2004 through 2008.[12] These data sources included state administrative data and newborn screening records, as well as case reports of patients from hemoglobinopathy specialty treatment centers. All administrative data sources used International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) codes, except for state death record files, which used International Classification of Diseases, 10th Revision (ICD-10) codes.[13,14]

Data collection: California
In California, data were requested from state newborn screening (all SCD patients born 2000 through 2008); vital records (state-based MCOD files, all deaths in 2004 through 2008); Office of Statewide Health Planning and Development: inpatient files (all records 2004 through 2008) and emergency department files (all records 2005 through 2008); Medi-Cal/Medicaid (all individuals insured in 2004 through 2008 who had an SCD ICD-9-CM code [282.41, 282.42, 282.6, 282.60, 282.61, 282.62, 282.63, 282.64, 282.68, 282.69]); hemoglobinopathy specialty treatment centers: Children's Hospital Los Angeles (all children and adults seen in 2004 through 2008 who had an SCD diagnosis) and University of California San Francisco Benioff Children's Hospital Oakland (all children and adults seen in 2004 through 2008 who had an SCD diagnosis).

After cleaning, data for patients found in multiple

datasets were linked by deterministic matching using social security number and date of birth where sufficient data were available, or by probabilistic algorithm[15] using weighted combinations of date of birth, sex, geographic area, facility attended, and diagnosis where sufficient data were unavailable. Details on data used, linkage, and results are available elsewhere.[12,16]

Data collection: Georgia
In Georgia, data were requested from state newborn screening (all SCD patients born 2004 through 2008); vital records (state-based MCOD files, all deaths in 2004 through 2008); Georgia Hospital Association: inpatient files (all records in 2004 through 2008) and emergency department files (all records in 2004 through 2008); Medicaid (all individuals insured in 2004 through 2008 who had an SCD ICD-9-CM code); Georgia's Children's Health Insurance Program (all individuals insured in 2004 through 2008 who had an SCD ICD-9-CM code); Georgia's State Health Benefit Plan (all individuals insured in 2004 through 2008 who had an SCD ICD-9-CM code); and hemoglobinopathy specialty treatment centers: Georgia Comprehensive Sickle Cell Center at Grady Memorial Hospital (all children and adults seen in 2004 through 2008 who had an SCD diagnosis), Georgia Health Sciences University (all children and adults seen in 2004 through 2008 who had an SCD diagnosis), and all three campuses of Children's Healthcare of Atlanta (all children seen in 2004 through 2008 who had an SCD diagnosis).

Few data sources included social security numbers or Medicaid identification numbers consistently; therefore, deterministic matching of patients was not feasible for most datasets in Georgia. Each dataset was de-duplicated and datasets were then matched, one at a time, using a probabilistic algorithm that assigned differing weights to identifying variables, such as date of birth, patient's name, mother's name (for children), sex, county, telephone number, and address. Details on data used, linkage, and results are available elsewhere.[12]

Inclusion criteria
This study included patients (referred to as SCD patients) who were reported by newborn screening and/or one of the reporting hemoglobinopathy specialty treatment centers with a laboratory-confirmed diagnosis (n=6,263). Patients with a combination of two or more health-care encounters with SCD ICD-9-CM codes and one or more expected SCD treatments, procedures, or complications (n=5,880) were also included.[12,16] This study did not include individuals having one or more health-care encounters with an SCD ICD-9-CM code but no evidence of an expected SCD treatment, procedure, or complication (n=17,642).

Mortality analyses
All SCD patients were linked to state-based MCOD files for 2004 through 2008 by using social security numbers or full names and dates of birth. All-cause mortality rates for the SCD population were calculated as the number of SCD patients who died from 2004 through 2008 in a given age group divided by the total number of SCD patients in that age group during the same period. The result was then divided by five for an annual average. For some SCD patients, the linked death records did not include SCD as a cause of death but otherwise met the inclusion criteria and were included in these analyses. Conversely, deaths in the state-based MCOD files that included SCD as a cause of death, but did not fit our inclusion criteria, are described separately and were not included in calculations of mortality rates or age at death.

All-cause mortality rates for the African American population and the general population in Georgia and California for 2008 only were derived by using CDC's Wide-ranging Online Data for Epidemiologic Research (WONDER) MCOD queries.[17] Data on SCD-coded deaths were also queried from the NCHS MCOD database and the NCHS CM database (also via CDC WONDER) for California and Georgia for years 2004 through 2008, for comparison with published SCD mortality surveillance reports and methods.[10,11,17,18] NCHS MCOD files are related to the state-based MCOD death files; both sets of files are derived from death certificates, both allow for coding of up to 24 underlying and contributing causes of death, and the methodology for coding is consistent across all states. State-based MCOD files, however, have patient-level data, include some identifiers, and are only offered to researchers and public health agencies for analysis under specific circumstances. NCHS MCOD files are reported to the public as aggregated data. In contrast with NCHS and state-based MCOD files, NCHS CM data indicate only the reported underlying cause of death and not contributing causes. As with the NCHS MCOD files, NCHS CM data are reported publicly as aggregate counts and rates and are not reviewable as individual patients. Also of note, both the NCHS MCOD and CM reports suppress results when a cell size is <10; therefore, information for some age groups may be incomplete if fewer than 10 patients in that age group died during the period of interest. We used ICD-10 coding to identify patients with SCD from the NCHS CM and NCHS MCOD files included in this study (D57.0, D57.1, D57.2, D57.8).

We calculated all-cause mortality rates for the SCD population in the two states (deaths per 1,000 patients) by age group at time of death, all-cause mortality rates for the African American population (deaths per 1,000 population), and all-cause mortality rates for all populations (deaths per 1,000 population) from both states. We calculated mean, standard deviation (SD), and median age at death by sex and overall. The data analysis for by this study was generated using SAS® version 9.3.[19]

RESULTS

Of the 12,143 SCD patients identified through population-based surveillance in California and Georgia in 2004 through 2008 and who met the inclusion criteria for this study, 615 were linked to records in state-based MCOD files. Among these 615 records, 143 were patients with known genotype and 472 were patients with unknown genotype. Proportions among age groups and sex were similar across the two states. Among these 615 SCD deaths, 16% of deaths occurred before 25 years of age and 45% of deaths occurred after 44 years of age (Table).

All-cause mortality rates for the SCD population (1.5 deaths per 1,000 patients) were lower than the all-cause mortality rates for African Americans (2.8 deaths per

1,000 population) in the youngest age group but rose steadily compared with the all-cause African American rates thereafter, until the oldest age group (Figure 1). For SCD patients, the mean (SD) age of death was 43.4 years (16.8) for females (median: 43.5 years) and 40.8 years (17.0) for males (median: 41.9 years); the mean age of death for all patients was 42.2 years (16.9) and median age of death was 42.8 years.

Data from the NCHS CM files showed 297 patients in Georgia and California with an underlying cause of death of SCD in 2004 through 2008; data from the NCHS MCOD files showed 513 patients with an underlying or contributing cause of death of SCD in 2004 through 2008; and data from population-based surveillance showed 615 SCD deaths in 2004 through 2008 (Figure 2). Sub-analysis of the NCHS data by age group produced information on 258 deaths from the CM file and 482 deaths from the MCOD file; these smaller numbers resulted from the suppression of cells with fewer than 10 deaths. The proportion of SCD deaths in the older age groups identified through population-based surveillance (≥45 years of age: 276/615, 45%) was larger than that identified by NCHS data (CM files: 95/297, 32%; MCOD files: 205/513, 40%). Deaths among the oldest age groups (≥65 years) occurred in 53/615 (9%) of SCD patients identified through population-based surveillance, 0/297 (0%) in the same

Table. Number of deaths for sickle cell disease patients identified using 2004–2008 population-based surveillance data, California and Georgia

| Characteristic | California and Georgia combined | | California | | Georgia | | Combined mortality rate per 1,000 patients per year |
	Number of SCD patients	Number of SCD deaths (percent)[a]	Number of SCD patients	Number of SCD deaths (percent)[a]	Number of SCD patients	Number of SCD deaths (percent)[a]	
Total	12,143	615	5,135	327	7,008	288	10.1
Age group (in years)							
0–4	1,738	13 (2)	746	8 (2)	992	5 (2)	1.5
5–14	2,686	18 (3)	1,096	10 (3)	1,590	8 (3)	1.3
15–24	2,435	68 (11)	979	36 (11)	1,456	32 (11)	5.6
25–34	1,986	101 (16)	781	46 (14)	1,205	55 (19)	10.2
35–44	1,496	139 (23)	656	72 (22)	840	67 (23)	18.6
45–54	1,097	142 (23)	524	79 (24)	573	63 (22)	25.9
55–64	474	81 (13)	236	45 (14)	238	36 (13)	34.2
65–74	164	32 (5)	83	19 (6)	81	13 (4)	39.0
≥75	67	21 (3)	34	12 (4)	33	9 (3)	62.7
Sex[b]							
Male	5,512	289 (47)	2,337	152 (46)	3,175	137 (48)	10.5
Female	6,630	325 (53)	2,798	175 (54)	3,832	150 (52)	9.8

[a]Column percentages may not total to 100 because of rounding.

[b]One Georgia patient was missing information on sex.

SCD = sickle cell disease

Figure 1. All-cause mortality rates for the sickle cell disease population identified through population-based surveillance (2004 through 2008), the African American population (2008),[a] and all populations (2008),[a] California and Georgia
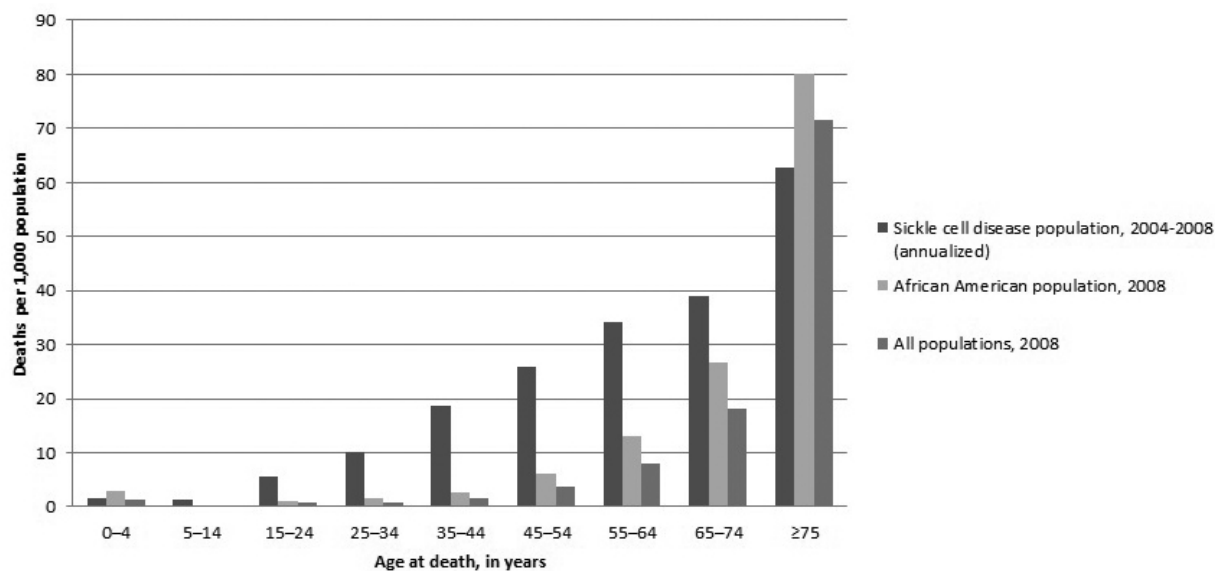
[a]Centers for Disease Control and Prevention (US), National Center for Health Statistics. Multiple cause of death 1999–2013 [cited 2015 Dec 15]. Available from: http://wonder.cdc.gov/mcd.html

age group in the NCHS CM files, and 13/513 (3%) in the NCHS MCOD data.

Among the 615 SCD patients linked to state-based MCOD data, 352 (57%) had an underlying or contributing cause of death coded as SCD, and the remaining 263 (43%) did not have SCD listed as a cause of death (Figure 3). In addition to the 615 state-based MCOD death certificates linked to SCD patients, 155 deaths were found in the state-based MCOD data (84 in California, 71 in Georgia) with an SCD ICD-10 code (excluding the code for sickle cell trait, D57.3), but no other evidence of SCD in the linked data sources. Among these 155 deaths were 81 deaths with an SCD code as the underlying cause of death and 74 with an SCD code as a contributing (but not underlying) cause of death. This group had a mean (SD) age of death of 45.0 years (17.8) and median age of death of 46.4 years; however, these 155 patients were not included in the analyses because they did not meet the inclusion criteria for our study.

DISCUSSION

This study was the first population-based SCD mortality study in the United States in which the denominator of patients with SCD was estimated from multiple and diverse linked data sources.[12] Our mortality rates were similar to previous reports from the United States,

Jamaica, and United Kingdom.[4,7,8,10,20–24] Our methodology for describing mortality in SCD identified more deceased patients and a higher proportion of patients living longer than did the methodologies in previous reports.[10,11] Our study identified 615 deaths among SCD patients in California and Georgia in 2004 through 2008. Comparable NCHS CM data yielded 297 SCD deaths during the same five years (48% of the deceased SCD patients), whereas NCHS's MCOD data yielded 83% of the deceased patient count. More importantly for understanding SCD outcomes, we found the all-cause mortality rate for the SCD population aged 5–74 years to be noticeably higher than all-cause mortality rate for the African American population and the all-cause mortality rate for the general population of the same age. We also found a higher proportion of SCD patients dying in the older age groups than did previous research.

Methodologically, our approach had several strengths over the previous research on SCD mortality. First, certain biases may be introduced when estimating survival from clinic-based populations, even when studied prospectively.[2,3] Late entry, attrition, selection of a potentially more symptomatic population of patients, and better medical management are among a number of limitations of this approach. Because we were able to analyze SCD mortality using a population-based sample of patients with SCD, our surveillance effort included

not only the fraction of the patient population being cared for by SCD experts, but also those who had care outside of sickle cell centers or were without medical homes or regular care providers.

Another challenge for previous studies was estimating the underlying prevalence of SCD in the population. Some researchers estimated it by using newborn screening data or by basing it on a portion of the total African American population.[4,5] However, these methods may not account for those not identified by newborn screening or early mortality in patients with SCD. Newborn screening was not instituted until the 1970s and did not become widespread in the United States until the late 1980s,[3] raising the potential for the older population of patients to be underrepresented in previous studies. Estimates based on newborn screening data also do not include those who were born outside of the United States.

Limitations

Although our study attempted to address many of the limitations of previous reports, several limitations remained. Only 23% of deceased SCD patients had a laboratory-confirmed diagnosis of SCD and were followed in comprehensive SCD programs or identified via newborn screening; however, we believe the remaining deceased individuals had a high probability of also being true SCD patients. These 472 patients without a laboratory-confirmed diagnosis were included as SCD patients based on multiple SCD-coded encounters in administrative datasets and relevant treatments, procedures, or complications that support the diagnosis; nevertheless, they did not have a known genotype, which limited the ability to determine mean and median ages at death by disease type. They may also differ significantly from the 143 SCD patients among whom the SCD genotype was confirmed because of the

Figure 2. Number of deaths for sickle cell disease patients identified using NCHS's compressed mortality reports (2004–2008),[a] NCHS's multiple cause of death reports (2004–2008),[b] and population-based surveillance data (2004–2008), California and Georgia
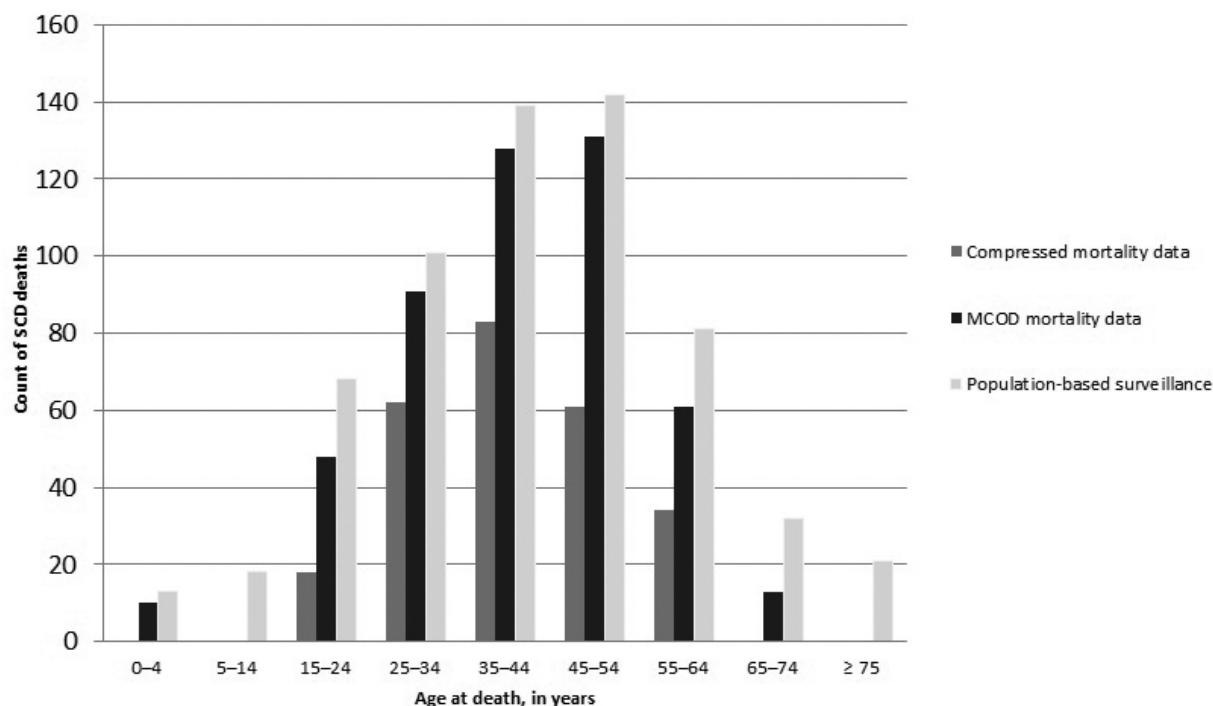
[a]Centers for Disease Control and Prevention (US), National Center for Health Statistics. Multiple cause of death 1999–2013 [cited 2015 Dec 15]. Available from: http://wonder.cdc.gov/mcd.html
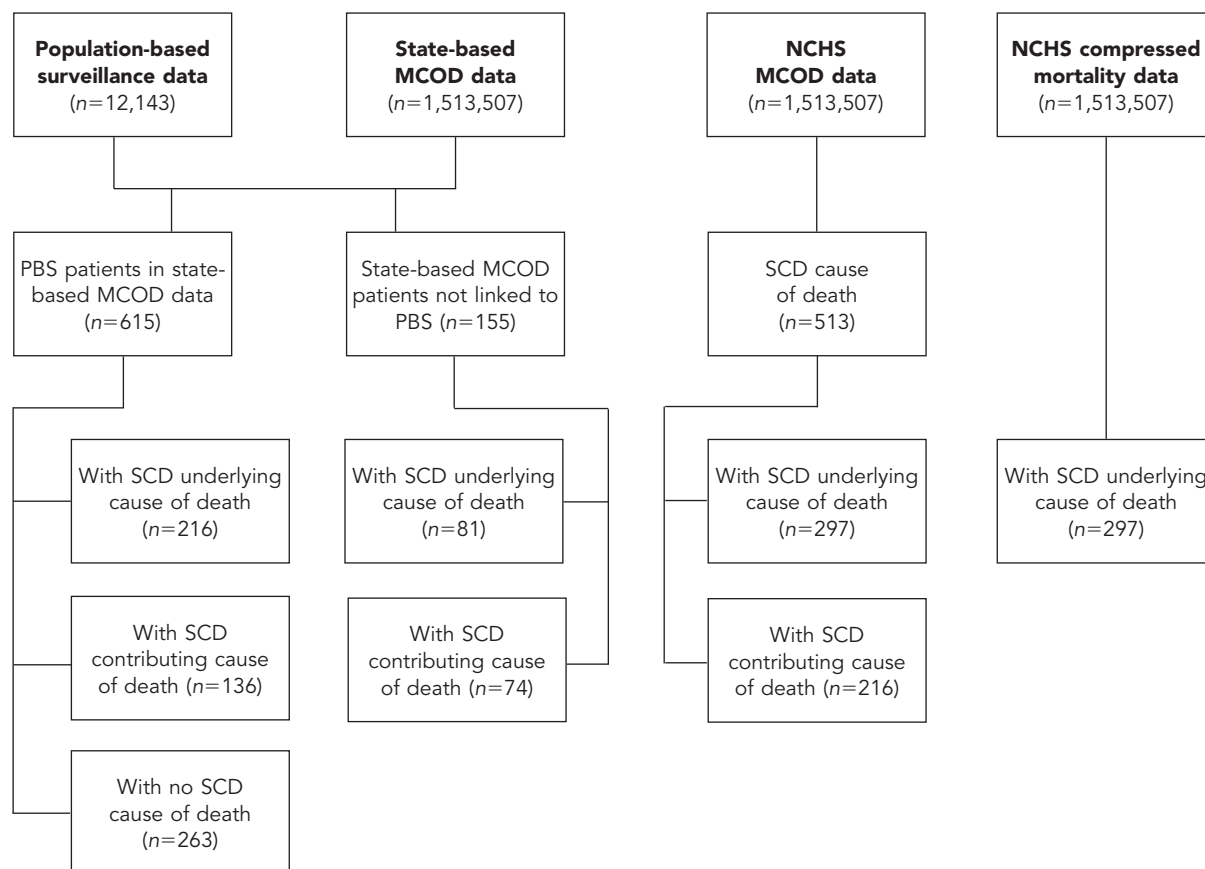
[b]Centers for Disease Control and Prevention (US), National Center for Health Statistics. Compressed mortality file 1999–2013 [cited 2015 Dec 15]. Available from: http://wonder.cdc.gov/cmf-icd10.html

NCHS = National Center for Health Statistics

MCOD = multiple cause of death

SCD = sickle cell disease

Figure 3. Number of deaths for sickle cell disease patients identified using population-based surveillance data (2004–2008), state-based multiple cause of death (MCOD) data files (2004–2008), NCHS's multiple cause of death reports (2004–2008),[a] and NCHS's compressed mortality reports (2004–2008),[b] California and Georgia

[a]Centers for Disease Control and Prevention (US), National Center for Health Statistics. Multiple cause of death 1999–2013 [cited 2015 Dec 15]. Available from: http://wonder.cdc.gov/mcd.html

[b]Centers for Disease Control and Prevention (US), National Center for Health Statistics. Compressed mortality file 1999–2013 [cited 2015 Dec 15]. Available from: http://wonder.cdc.gov/cmf-icd10.html

PBS = population-based surveillance

MCOD = multiple cause of death

NCHS = National Center for Health Statistics

SCD = sickle cell disease

underlying differences in the ways these two groups were identified. The patients with confirmed genotypes were from newborn screening data or from specialty SCD programs, so they were likely younger than the general SCD population, and/or received comprehensive care, and/or were referred to a specialty center because of the severity of their disease.

Furthermore, although validation efforts used in the Registry and Surveillance System for Hemoglobinopathies study suggest a small error rate in using this approach of combining diagnosis and complications to identify patients,[25] and our results were similar to

other studies that examined the accuracy of using diagnosis codes for SCD patient finding,[26] patients who were older and/or living with milder disease may be underrepresented in these data. Consequently, the normalization of the mortality rate among the oldest group of SCD patients may result from the underlying patient population's small size (n=67), a high proportion of older patients who did not actually have SCD, yet met our study's inclusion criteria, and/or an older population more likely to have milder disease and more likely to die from non-SCD-related causes. Furthermore, our methods did not allow us to determine if

identified patients moved out of California or Georgia during the five-year period and who, if so, should not have been included in the denominator for mortality calculations. Additionally, it is impossible to derive measures of central tendency from NCHS data (either the MCOD or CM files) because deaths are aggregated by age group; we therefore compared the means and medians reported in the earlier studies that are from earlier years of collected data and not specific to only these two states.[4,5] Finally, this analysis includes only the 5% of the Registry and Surveillance System for Hemoglobinopathies patients who had died during a five-year period of observation from 2004 through 2008.

CONCLUSION

Surveillance data from the Registry and Surveillance System for Hemoglobinopathies project provides the most accurate data on mortality among the sickle cell population reported to date. Although SCD was once considered nearly universally fatal by adolescence, many patients are now living well into adulthood[6] and surviving to 60 years of age or older.[3] Documenting complications and outcomes during the life course informs development of standards of care, interventions, and health-care policy to serve this population. It also highlights the importance of routine health screening for malignancies, diabetes, heart disease, and other non-SCD conditions common among older adults. Accurate assessment of life expectancy, causes of death, and disease prognosis requires long-term surveillance using population-based registries of patients with SCD accurately diagnosed at birth.

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