Colorado Sickle Cell Data Collection Program: Surveillance and Prevalence of Sickle Cell Disease in Colorado

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Table of Contents

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Abstract

Background: Sickle cell disease (SCD) is a hereditary blood disorder causing significant health complications. While advancements in care have improved survival rates, accurate prevalence data in Colorado has been limited, hampering effective public health interventions.

Objective: The objectives of this study were to establish and describe the Colorado Sickle Cell Data Collection (CO-SCDC) program, update the prevalence and birth prevalence of sickle cell disease (SCD) in Colorado, and improve surveillance methodologies to inform public health interventions, education initiatives, and policy development.

Methods: The CO-SCDC program collected and linked data from multiple sources including the Colorado Department of Health and Environment and electronic health records from University of Colorado Health and Children's Hospital Colorado. Data from 2012 to 2020 were analyzed using national case definitions to classify SCD cases. Case validation was performed via clinical review by two hematologists. Prevalence and birth prevalence of SCD in Colorado were calculated as were demographics of the Colorado SCD population.

Results: In 2019, 435 individuals were identified as living with SCD in Colorado, an increase of 16-40% over previous estimates. The prevalence of SCD was highest in urban counties, with concentrations in Arapahoe, Denver, and El Paso counties. Birth prevalence of SCD increased from 11.9 per 100,000 live births between 2010-2014 to 20.1 per 100,000 live births between 2015-2019. The study highlighted a 67% increase in SCD births over the decade, correlating with the growth of the Black or African American population in the state.

Conclusions: The CO-SCDC program successfully established a capacity to perform SCD surveillance and a baseline of prevalence estimates for SCD in Colorado. The findings underscore the importance of surveillance for informed public health strategies and policy development to improve outcomes for individuals with SCD.

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Keywords: sickle cell disease, public health surveillance, prevalence, Colorado

Abstract

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Results: In 2019, 435 individuals were identified as living with SCD in Colorado, an increase of 16-40% over previous estimates. The prevalence of SCD was highest in urban counties, with concentrations in Arapahoe, Denver, and El Paso counties. Birth prevalence of SCD increased from 11.9 per 100,000 live births between 2010-2014 to 20.1 per 100,000 live births between 2015-2019. The study highlighted a 67% increase in SCD births over the decade, correlating with the growth of the Black or African American population in the state.

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Introduction

Sickle cell disease (SCD) is a hereditary blood disorder characterized by the presence of abnormal red blood cells that assume a rigid, sickle-shaped form, leading to impaired blood flow and various health complications.[1] Recent advancements in healthcare have significantly improved the survival rates of individuals with SCD, with over 95% of patients born in developed countries expected to survive into adulthood due to enhanced supportive and preventive care measures such as newborn screening, penicillin prophylaxis, and transcranial Doppler screening.[2] Despite improvements, the disease still poses significant challenges, with potential worsening of complications in adolescence

and young adulthood, potentially compounded by reduced adherence to treatment protocols and the need to transition from pediatric to adult care.[3] Mortality rates among adults with SCD have increased over time, possibly indicating limited access to and/or underutilization of high-quality care for this demographic.[4]

Estimates suggest that the number of individuals affected by SCD in the United States (US) could be 100,000, though limitations in data quality and variations among states hinder precise estimations.[5] In 2010, Hassell [6] published SCD population estimates for the US derived from the 2008 United States Census and the birth-cohort disease prevalence from the National Newborn Screening Information System (NNSIS). Hassell estimated a NNSIS-based total birth cohort between 311 and 371 individuals with SCD living in Colorado (CO) between 2005 and 2007, after correcting for early mortality. These prevalence estimates for CO have not been updated since.

The Centers for Disease Control and Prevention (CDC) initiated the Registry and Surveillance System for Hemoglobinopathies (RuSH) project [7],[8] in 2010 to enhance estimates of SCD. This marked the inception of the first comprehensive surveillance initiative for SCD in the United States, incorporating data from multiple sources. Subsequently, the Public Health Research, Epidemiology, and Surveillance for Hemoglobinopathies (PHRESH) project [7, 8], conducted from 2012 to 2014, aimed to expand the knowledge derived from RuSH data, refining surveillance methodologies, and instituting strategies for health promotion and the prevention of associated health complications.

The Sickle Cell Data Collection (SCDC) program subsequently began in 2015 to expand SCD surveillance activities by collecting diagnosis, treatment, and health care utilization data on individuals with SCD. Colorado was selected for funding in 2021 and established the Colorado SCDC program (CO-SCDC) with the aim of establishing a SCD surveillance system to serve the CO SCD community by providing relevant data that would inform public health interventions, education initiatives, and policy development.

In this manuscript, we describe the CO-SCDC project, the collaborative partnerships developed in the state and the methods used to obtain SCD surveillance data within CO. We report updated SCD population prevalence and birth prevalence numbers based on the results of public health surveillance methodologies that collate multiple large-scale data sources across the state.

Methods

The CO-SCDC Collaborative Team

The Center for Public Health Innovation (CPHI) established a multidisciplinary team (MDT) across multiple institutions in 2021 to manage the CO-SCDC program. The MDT is comprised of

epidemiologists, health data analysts, program evaluators, and health communication experts at CPHI, and adult and pediatric hematologists at the Colorado Sickle Cell Treatment and Research Center (CSCTRC) at the University of Colorado School of Medicine (CU Medicine) who are involved with the clinical services provided to individuals with SCD at the Children's Hospital Colorado (CHCO) and the University of Colorado Health (UCHealth) systems. The MDT also consists of leaders from the Colorado Sickle Cell Association (local community-based organization), representatives from newborn screening and vital statistics programs in the Colorado Department of Public Health and Environment (CDPHE), a chronic disease surveillance specialist from the Colorado School of Public Health, a mental health professional specializing in pain management at Road to Me Recovery, an employment services expert at the Colorado Department of Vocational Rehabilitation, and a SCD transition specialist social worker from the CSCTRC.

Data Acquisition

CO-SCDC acquired linked and de-identified data from Health Data Compass (Compass), the honest broker for the project. Compass is a multi-institutional data warehouse funded by UCHealth, CHCO, and CU Medicine, and is designed to support data discovery and data science methodologies that integrate, harmonize, and link large-scale biological, clinical, administrative, regional, state, and national data sets. Compass incorporates electronic health record (EHR) data from all health records within the UCHealth and CHCO systems across CO. This includes records from individuals seen across four CHCO hospitals and six outpatient clinics, and twelve UCHealth hospitals and over 120 outpatient clinics in CO. Compass also receives notification of deaths in the state of Colorado for all individuals in the Compass system through a partnership with the CO Vital Statistics Program.

Healthcare encounters across all CHCO and UCHealth hospitals and outpatient clinics for individuals with SCD-related International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) [9] and ICD-10-CM [10] codes from January 1, 2012, through December 31, 2020, were pulled by Compass and deidentified.

The CO NBS program provided data from January 1, 2001, to December 31, 2020, on infants with a confirmed SCD diagnosis. These data included date of birth, biological sex, mother's basic demographics, and final diagnostic SCD sub-type. To obtain race, ethnicity, and county of residence at birth, NBS data was merged with birth records through the CO Vital Statistics Program. The Vital Statistics Program linked the NBS records with the specific birth record fields of interest. Once linked, the NBS data and birth record data were entered into a REDCap database housed at UCHealth.[11],[12] The individual-level, fully identifiable data were pulled by Compass and linked with patient data for individuals with UCHealth and CHCO healthcare encounters and SCD-related

ICD-9-CM and ICD-10-CM codes.

All data linkages at Compass were performed by utilizing the Colorado University Record Linkage (CURL) tool to match and merge records for all data listed above. CURL is a platform that performs record linkage operations and uses hashing to encode sensitive data into random strings of information ensuring that patient data is secure across all parties involved once the data is linked.[13] All linked identifiable data were stored on secure servers at UCHealth Compass. Data was then de-identified and transferred as limited datasets directly to the CPHI Microsoft Office E5 level HIPAA compliant OneDrive.[14]

Analysts at CPHI cleaned and formatted the data and performed additional de-duplication. Demographic fields, dates, diagnostic data, and acute care utilization fields were reformatted to match across all institutions. Potential data errors were identified and corrected or removed. Arbitrary person identifiers assigned by Compass were compared within and across institutions and deduplicated. Any healthcare encounters with a county of residence outside of CO were excluded. All data processes were performed using SAS version 9.4 (SAS Institute Inc).

Case Definitions

After data cleaning, we applied national SCDC case definitions to determine which individuals represented a case of possible, probable, or definite SCD.[7, 15].[16] Possible cases were identified as any individual with three or more healthcare encounters across participating hospitals, emergency departments, or outpatient clinics with a SCD ICD-9-CM or ICD-10-CM code over any 5-year period between January 1, 2012 and December 31, 2019. Possible cases were upgraded to probable if the individual had a hospital discharge in 2019, emergency department visit in 2019, or both before and after 2019. A case was confirmed as definite if they had a confirmed diagnosis of SCD after a presumptive positive newborn screen or were confirmed to have SCD via chart review by a clinical hematologist at the CSCTRC. Only probable and definite cases were reported.

Case Validation

All possible, probable, and definite cases from the year 2018 were submitted to one adult and one pediatric hematologist at the CSCTRC for case validation. The hematologists went through the electronic health record and determined whether an individual was a definite case, not a case, or unsure if not enough information was available. All individuals determined to be a definite case were included in the counts for 2019. All individuals determined not to have SCD were removed from the cohort and excluded from the analysis, as were individuals determined to have sickle cell trait. Individuals with a bone marrow transplant (BMT) prior to 2019 were considered cured and excluded.

Analysis

Frequencies and descriptive statistics were computed for both probable and definite cases. Positive predictive values (PPV) were calculated by dividing the true positives (TP), or those confirmed to have SCD, by the total number of cases found using the case definitions. Negative Predictive Values (NPV) were calculated by dividing the true negatives (TN), or those confirmed not to have SCD, by the total number of negative cases found using the case definitions. Prevalence rates at the state and county levels were determined by dividing the combined number of definite and probable cases by the estimated population of individuals residing in CO or specific CO counties, as per the 2020 US Census population estimates.[17] These rates were then multiplied by 100,000 to obtain the prevalence per 100,000 individuals. Prevalence numbers by race were calculated similarly by using the total Black or African American population as the denominator. Birth prevalence rates were calculated by dividing the number of confirmed SCD births in CO by the total number of live births in CO[18] and then multiplied by 100,000 to obtain birth prevalence rates per 100,000 people. Birth prevalence by race was calculated similarly by using the total Black or African American live births as the denominator. All data analyses were performed using SAS version 9.4 (SAS Institute Inc).

Results

2018 SCD Case Validation

There was a total of 1,374 individuals with a SCD ICD-9-CM or ICD-10-CM code identified during the years 2012 through 2020. After excluding encounters from individuals who were not CO residents, deceased, only had encounters in 2019-2020, had an ICD-9-CM or ICD-10-CM code for a BMT, and applying case definitions, 566 (58.8%) met the possible, probable, or definite case definition criteria for the year 2018 and were presented for clinical validation (Figure 1). A total of 312 were probable or definite cases and therefore considered positive cases through the application of the case definitions for the year 2018. Of these, 295 were identified as true positives with a PPV of 94.6%. A total of 254 possible surveillance cases were also validated and combined with 396 individuals who did not meet any level of case definition criteria for a total of 650 negative cases. Of these, 513 were identified as true negatives with a negative predictive value of 78.9%. The remaining 137 false negative cases were then classified as positive cases and added to the 295 true positive cases. These 432 cases were indexed and carried over as true cases for 2019 analyses. *[insert Figure 1]*

2019 Surveillance Cases

Of the 432 cases carried over as validated cases for 2019, there were three deaths in 2019, four with

insufficient data for reporting purposes, and eight individuals who were identified via case validation to have had a BMT in 2019 or prior not identified via ICD code, leaving 417 validated definite cases from the 2018 dataset. After applying the specified case definitions to the 2019 surveillance data, an additional 18 cases met probable or definite criteria for SCD in CO, bringing the 2019 total to 435 cases (Figure 1). Most of these individuals were identified as Black or African American (80.2%) and non-Hispanic (93.3%). The median age within this population was 19, with an interquartile range (IQR) of 8 to 30 (Table 1).

Table 1. Demographics of Individuals with Sickle Cell Disease in Colorado, 2019.

Demographic	**N (%)**
Total	435 (100)
Sex	
Male	215 (49.4)
Female	220 (50.6)
Age (Years)	
<10	119 (27.4)
10-19	100 (23)
20-29	96 (22.1)
30-39	78 (18)
40-49	27 (6.2)
50+	15 (3.4)
Race	
Black or African American	349 (80.2)
Other	75 (17.2)
Unknown	11 (2.5)
Ethnicity	
Hispanic	18 (4.1)
Non-Hispanic	406 (93.3)
Unknown	11 (2.5)

2019 SCD Geographic Prevalence

Most individuals lived in urban counties (97.5%), but geographic distribution also spread to less populated counties on the western slope and eastern rural areas of CO (Figure 2). The three counties with the highest prevalence rates of SCD were Arapahoe (18.8 cases per 100,000 people), Denver (16.2 cases per 100,000 people), and El Paso (11 cases per 100,000 people). The county with the highest rate of SCD by race or ethnicity was Adams with 230.2 cases per 100,000 Black or African American individuals, then Denver County with 149.8 cases per 100,000 Black or African American individuals, and finally Arapahoe with 142.1 cases per 100,000 Black or African Americans (Figure 2).

2015 to 2019 SCD Birth Prevalence

There were 195 SCD births in CO between 2000 and 2019. The number of births during a 5-year period increased from 39 births between 2010-2014 to 65 births between 2015-2019 (Figure 3). The birth prevalence of SCD in CO during a 5-year period also increased from 11.9 per 100,000 live births between 2010-2014 to 20.1 per 100,000 live births between 2015-2019, equivalent to 1 in 4,974 live births. When categorized by race and ethnicity, the birth prevalence among Black or African American individuals increased from 221.9 per 100,000 live births to 331 per 100,000 live births between 2015-2019, or 1 in 302 live births among this demographic. Congruently, the number of Black or African American live births during a five-year period increased by 32.8% between the periods of 2005-2009 and 2015-2019, in contrast, the total number of all live births in CO decreased by 4.5% between these same five-year periods (Figure 4). Between 2015 and 2019, most SCD births were of the subtype HbSS or HbSβ^0 thalassemia (n = 47, 58.8%), followed by HbSC (n = 19, 23.8%) and HbSβ^+ thalassemia (n = 11, 13.8%).

Discussion

Historical Estimates and Current Findings

Previously published estimates from the Agency for Health Care Policy Research estimated that approximately 640 people with SCD lived in CO as of 1993.[19] The National Newborn Screening Information System (NNSIS) used birth cohort data to estimate that between 311 and 371 individuals with SCD lived in CO between 2005 and 2007, after correcting for early mortality.[6] The CO-SCDC project found 435 individuals living with SCD in CO in 2019. Methods used for the CO-SCDC project resulted in higher counts than the NNSIS estimate by between 17% and 40%. This could be attributed to the 27.4% increase (68,367 individuals) of individuals in the Black or African American community in CO since 2010.[17],[20] The increase in birth prevalence supports the population growth theory as SCD births in a 5-year period increased by 67% between 2010 and 2019, likely due to the increase of the Black or African American population in CO and the 11.7% increase in the birth rate among this population in a 5-year period between 2010 and 2019. Additionally, advancements in surveillance techniques and data methodologies employed for the project likely contributed to more accurate SCD estimates within the population.

Accessibility and Distribution of Care

The state of Colorado is home to a comprehensive SCD center, the CSCTRC, located near the epicenter of the Denver metropolitan area where approximately 70% of individuals living with SCD in Colorado reside (Figure 2). Sickle Cell Center-affiliated providers serve patients within the major

healthcare systems serving both children and adults in this area and provide care at an outreach clinic in the central mountain area. CSCTRC-affiliated specialty practices that see SCD patients are also located in Fort Collins and Colorado Springs, CO. This increases the coverage in the Front Range of CO to approximately 90% of individuals with SCD living in CO within a 60-mile radius of the healthcare systems with dedicated sickle cell specialists or affiliated specialty practices. This means around 10% of individuals with SCD in CO reside in counties on the Western Slope, Eastern Plains, or Southern CO where specialty SCD care is not readily accessible. This is important to note, as patients in these locations often require assistance to locate appropriate care facilities and to secure a provider with specialized expertise in SCD, particularly during the transition from pediatric to adult care. These factors, compounded by the scarcity of providers in rural regions willing to see sickle cell patients, are among the most frequently reported barriers to access to care among individuals with SCD.[21] The availability of surveillance data to identify areas of need informs the work of ongoing programs, such as the statewide Sickle Cell Center Transition Program, to correct these gaps.

Surveillance and Data Methodologies

The case validation process proved crucial to the surveillance case identification process for the CO-SCDC project. Though the case definitions were very proficient at identifying true clinical cases with a PPV of 95.6%, additional validation was required as 53.9% of possible cases were confirmed via clinical review as definite SCD cases. Had we not performed the case validation, the official surveillance estimates of SCD in 2019 would have underestimated the total count by 137 individuals (31.5%). This emphasizes the importance of manual case identification in combination with automated data processes to arrive at the most accurate results possible. While this proved a valuable and attainable goal in CO, manual case review may prove more burdensome in states with higher SCD prevalence.

Implications for Policy and Community Programs

The significance of utilizing this data to implement change in the community cannot be overstated. The CO Sickle Cell Association (CSCA), the local community-based organization, plays an instrumental role in this project in disseminating and leveraging the data to tailor education and policy initiatives to address specific challenges. SCD counts reported in this manuscript were recently used by the CSCA to advocate before the CO General Assembly for a bill that was passed to establish an SCD outreach program to provide support to individuals living in CO with SCD.[22] Looking ahead, it is important that this data continue to expand and refine, helping the CSCA to further bridge the gap between healthcare providers, policymakers and the sickle cell community.

Limitations

This project had several limitations. Data received from Compass includes the UCHealth and CHCO network centers, hospitals, and outpatient clinics which collectively encompass the comprehensive SCD treatment center and affiliated specialty practices but doesn't include healthcare encounters from providers in the state outside of these networks. Also, access to specific state Medicaid administrative data was not available in CO. Future analyses will incorporate all-payer claims data from the Center for Improving Value in Health Care to ensure the project is as comprehensive as possible by including data from healthcare encounters outside the CHCO and UCHealth networks. Additionally, there were complexities with identifying whether a SCD case still resided in CO for the reporting year and project analysts will need to find an algorithmic solution moving forward.

Acknowledgments

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Conflicts of Interest

None declared.

Abbreviations

BMT: bone marrow transplant
CDC: Centers for Disease Control and Prevention
CDPHE: Colorado Department of Public Health and Environment
CHCO: Children's Hospital of Colorado
CO: Colorado
CO-SCDC: Colorado Sickle Cell Data Collection Program

Compass: Health Data Compass
CPHI: The Center for Public Health Innovation
CSCA: Colorado Sickle Cell Association
CSCTRC: Colorado Sickle Cell Treatment and Research Center
CU Medicine: University of Colorado School of Medicine
CURL: Colorado University record linkage
EHR: electronic health record
ICD-10: International Classification of Diseases, Tenth Revision
ICD-9: International Classification of Diseases, Ninth Revision
MDT: multidisciplinary team
NNSIS: National Newborn Screening Information System
NPV: negative predictive value
PHRESH: Public Health Research, Epidemiology, and Surveillance for Hemoglobinopathies
PPV: positive predictive value

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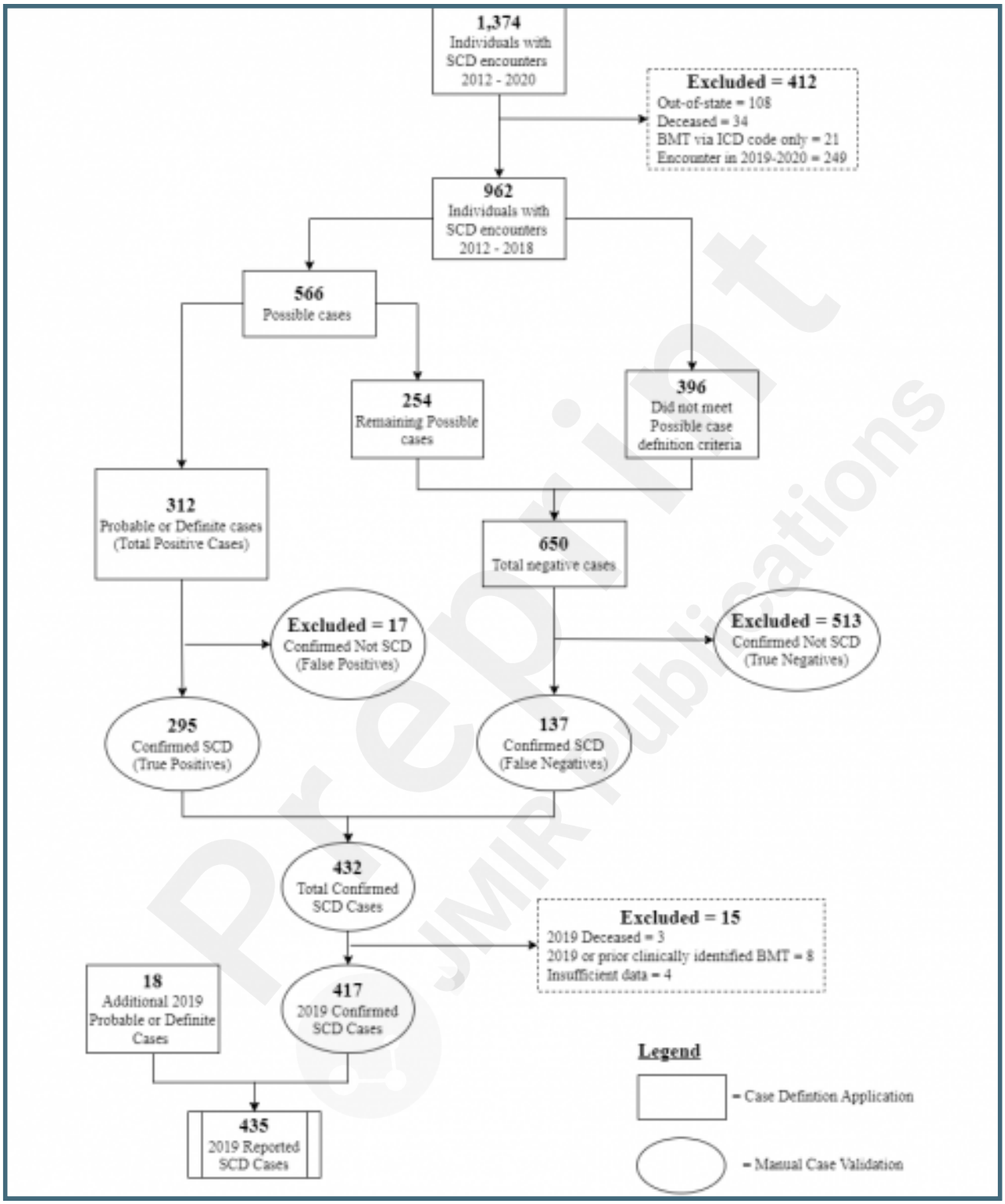
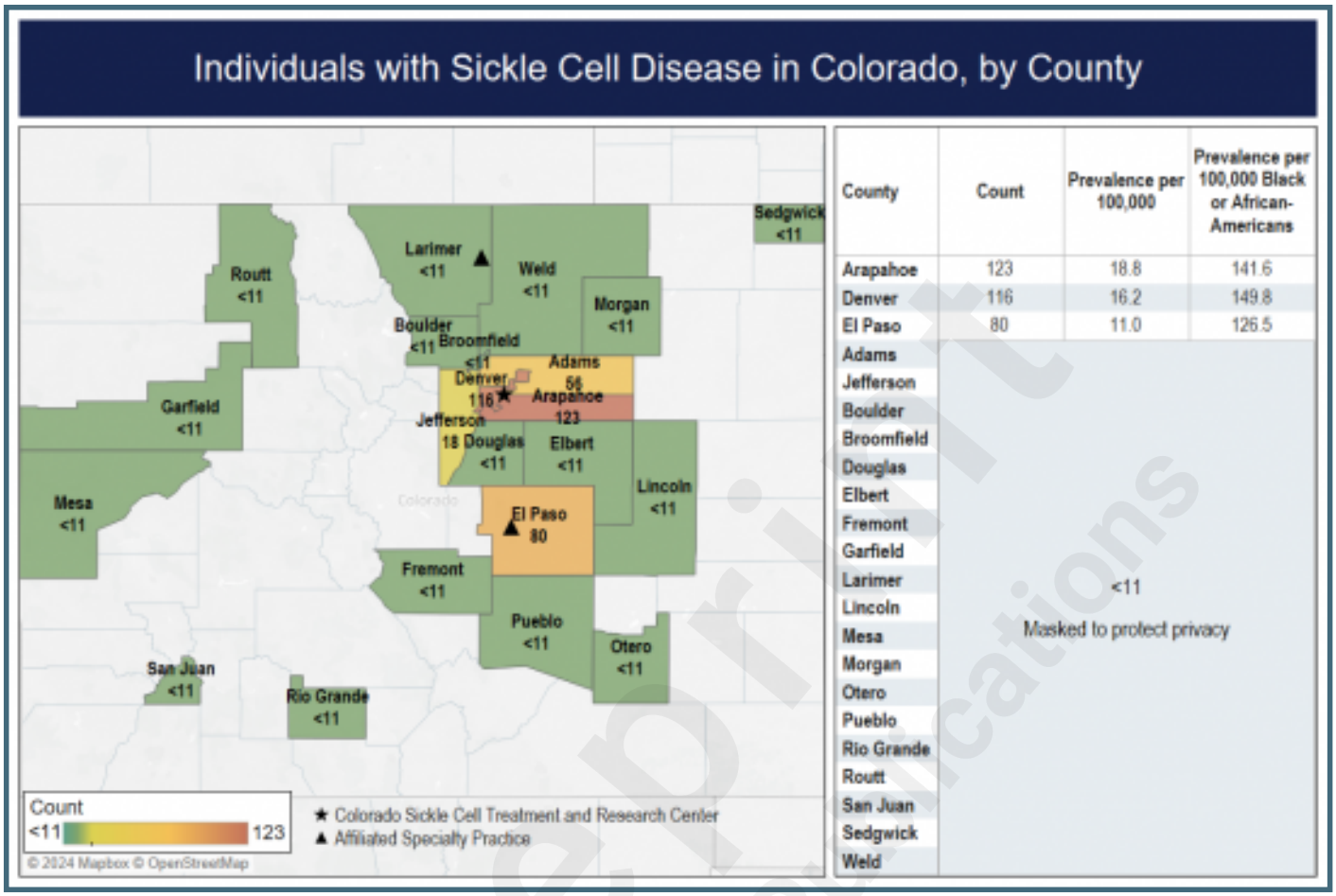
Supplementary Files

Figures

CONSORT diagram showing 2018 case validation and 2019 case determination flow.

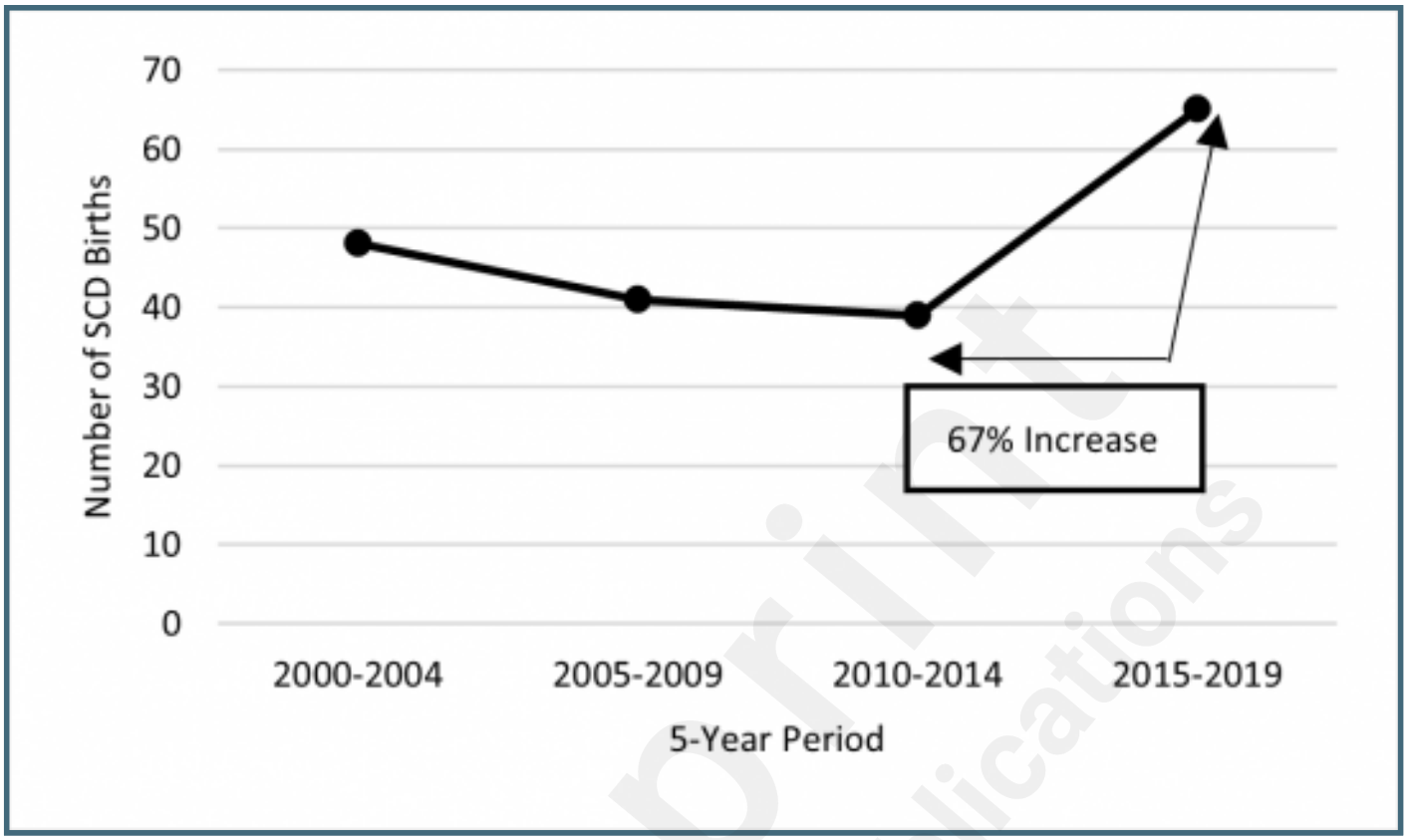


Colorado sickle cell disease counts and prevalence by county, 2019.



County	Count	Prevalence per 100,000	Prevalence per 100,000 Black or African-Americans
Arapahoe	123	18.8	141.6
Denver	116	16.2	149.8
El Paso	80	11.0	126.5
Adams	<11	Masked to protect privacy	
Jefferson			
Boulder			
Broomfield			
Douglas			
Elbert			
Fremont			
Garfield			
Larimer			
Lincoln			
Mesa			
Morgan			
Otero			
Pueblo			
Rio Grande			
Routt			
San Juan			
Sedgwick			
Weld			

Then number of Colorado sickle cell births in a 5-year period increased by 67% between 2010 - 2019.



The number of Colorado black or African American births in a 5-year period increased by 32.8% between 2000 - 2019 while all births decreased by 4.5%.