

California Sickle Cell Action Plan 2018 Executive Summary



Prepared by California Sickle Cell Stakeholders under the leadership of the Pacific Sickle Cell Regional Collaborative. For more information visit pacificscd.org or casicklecell.org.

Background

Sickle cell disease (SCD) is a painful, rare, progressively debilitating inherited blood disorder that has been unrecognized for decades in the U.S., leading to significantly poor health outcome and resource inequities. **Sickle cell trait (SCT)** is not a disease, but having it means that a person has inherited the sickle cell gene from one of their parents. People with SCT usually do not have any of the symptoms of SCD, but they can pass the sickle cell gene on to their children.¹

The situation is urgent in California, where preventable deaths are occurring, needlessly shortening the lives of adults with SCD. The California Sickle Cell Action Plan (CA-SCAP) was informed by diverse California stakeholders concerned about the lack of access to comprehensive, evidence-informed care for individuals living with SCD in the state. This Executive Summary presents our work to identify the issues and to outline strategies for improving sickle cell care in California.

Current State of Sickle Cell in California

The median age of death for individuals with SCD in California is 43 years, well below the U.S. general population, the overall African American population, and people with SCD in other U.S. states.

There now exists an almost perfect storm of factors that

make it hard for older teenagers and adult Californians with SCD to get comprehensive and appropriate SCD care. These factors include:

- 1) A lack of primary care providers knowledgeable about SCD and a dearth of specialists who treat adolescents and particularly adults with SCD;
- 2) Inadequate reimbursement for adult SCD treatment that is preventive and team-based;
- 3) Few adult SCD centers for comprehensive care;
- 4) Poorly planned and executed transition from pediatric to adult medical care.; and
- 5) A lack of knowledge on pain management techniques for individuals with SCD.

Social determinants of health, including poverty, lack of access to transportation, homelessness, cultural and language barriers, racism, and discrimination exacerbate the challenges children and adults with SCD face in accessing comprehensive care. Furthermore, the initiatives to address the current opioid crisis inadvertently limit access to treatment for debilitating SCD- related pain, wrongly stigmatizing people with SCD as drug seekers, resulting in the denial of appropriate care. This situation can and must be addressed.

“People with sickle cell disease are dying because of the lack of healthcare infrastructure in California”

Dr. Elliott Vichinsky

Director, Comprehensive Sickle Cell Center,
UCSF Benioff Children’s Hospital Oakland

California Sickle Cell Action Plan

The California Sickle Cell Action Plan (CA-SCAP) creates a roadmap forward, presenting long-term goals and strategies to improve healthcare systems, and increase education and awareness about sickle cell disease and sickle cell trait in California. Its ultimate aim is to improve the quality of life and longevity of those living with sickle cell disorders now and in the future. While our goals are ambitious, our priorities are clear.

First Priority: To work with key public and private partners to make sickle cell a priority in California with a synergistic call to action.

SCD in California primarily affects African American and Latino families, underserved populations who suffer added challenges when they have this complex disease that impacts every facet of their lives.

Other states have had focused multiagency efforts to raise SCD as a health priority to improve quality of life, appropriate health services utilization, cost savings, and to increase life expectancy.^{2,3}

This plan outlines a path for the California Department of Health Care Services (CDHCS), California Department of Public Health (CDPH) and other public and private entities to work together to create opportunities and remove barriers to evidence-informed care for people living with or at risk for SCD.

We expect that key partner agencies will devote resources to support implementation of this statewide sickle cell initiative within their own organization's scope/mission. Much of this work can be accomplished using existing public and private programs, frameworks, and plans.

Increasing education and awareness about SCD as a vital health equity concern in California and committing resources to eliminate disparities are key elements of this priority.



Second Priority: To increase health care services for Californians living with SCD by:

- 1. Establishing or strengthening regional centers of excellence** providing team-based, integrated care including pain management, mental health, trait testing and genetic counseling. Teams at these centers will also address the social determinants of health;
- 2. Increasing adequate funding** to cover reimbursement for clinical and non-clinical services as well as the establishment of adult SCD specialty clinics;
- 3. Identifying and implementing workforce development** in primary and specialty care, with a focus on SCD; and
- 4. Supporting long-term surveillance** of SCD complications, mortality, health services utilization, and cost throughout the lifespan to understand which populations are most vulnerable and identify emerging trends for action.

Goals and Strategies

Over 50 stakeholders from throughout the state met regularly in 2017-18 to generate goals and strategies to address sickle cell health inequities in California. We present here both short term and aspirational goals from our Community Needs, Clinical Care, and Policy workgroups, as synthesized by our steering committee.

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| Goal 1: PRIORITY | Prioritize Sickle Cell in the State of California |
| Strategy 1.1 | Ensure that sickle cell is named a CDPH and CDHCS priority population. |
| Strategy 1.2 | Stakeholders support the CA-SCAP by advocating with decision makers – legislators, state agencies including CDHCS, CDPH, GHPP, and CCS, and hospital and health care leaders – to support and implement CA-SCAP policy goals. |
| Goal 2: AWARENESS | Increase Sickle Cell Education and Awareness |
| Strategy 2.1 | <ol style="list-style-type: none"> 1) Increase SCT and SCD education. 2) Develop statewide multi-level messaging and communication strategies, including a focus on schools, to increase awareness of SCT and SCD, particularly among those who are at risk for having SCT and SCD. |
| Strategy 2.2 | Raise public awareness about SCT and SCD through strategic partnerships. |
| Strategy 2.3 | Use existing networks of advocates, community-based organizations, and clinical facilities to provide patient education to families living with SCD everywhere in the state. |
| Goal 3: CARE | Increase Health Care Services for Californians Living With Sickle Cell Disease |
| Aim 1: SERVICES | Address Gaps in Care |
| Strategy 3.1.1 | Improve reimbursement/payment systems to support sickle cell services for prevention, treatment, education and care coordination, and to address social determinants of health. |
| Strategy 3.1.2 | <ol style="list-style-type: none"> 1) Improve excellence in care in all settings, thereby improving patient experiences and outcomes in acute care settings. 2) Ensure that populations at risk have adequate access to diagnosis, education, and outreach, treatment (including primary and specialty care), rehabilitation, and care transitions for SCD and SCT across the lifespan. |
| Strategy 3.1.3 | Improve comprehensive pain management. |
| Strategy 3.1.4 | Increase access to mental health and psychosocial services. |

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| Strategy 3.1.5 | Increase access to, and utilization of SCT testing and genetic counseling. |
| Strategy 3.1.6 | Create SCD “hubs and spokes”* of clinical excellence** and eliminate barriers to them (Figure 1). |
| Strategy 3.1.7 | Address social determinants that impact health and health outcomes, such as transportation and housing, supporting Community Health Workers and Patient Navigators as essential members of sickle cell teams. |
| Strategy 3.1.8 | 1) Strengthen and act upon understanding of trends in sickle cell related complications, treatments, utilization, quality of life and life expectancy by public health surveillance. 2) Enhance and sustain system of public health surveillance and research for SCT and SCD. |
| Aim 2: WORKFORCE | Improve Sickle Cell Care Workforce Based on Expert Guidelines Across the Lifespan |
| Strategy 3.2.1 | Raise awareness and educate practicing and next generation health professionals about the needs of people living with SCD and their families, including trait testing and trait education through workforce development. |
| Strategy 3.2.2 | Ensure that a group of identified pediatric and adult primary care providers, trained in guidelines for SCD care, will provide primary care and coordinate SCD care with specialists. |
| Aim 3: TRANSITION | Assure Timely and Complete Transition to Adult Care |
| Strategy 3.3.1 | Increase access to knowledgeable, guideline-based care for adults with SCD. |

* The hub-and-spoke is a model which arranges service delivery assets into a network consisting of an anchor establishment (hub) which offers a full array of services, complemented by secondary establishments (spokes) which offer more limited service arrays, routing patients needing more intensive services to the hub for treatment.⁴

** Center of Clinical Excellence: A diagnostic and treatment center that provides multidisciplinary care, which consistently and persistently sees a large enough volume of patients with sickle cell disease to do the following:

- 1) Develop and maintain expertise and resources to provide the care patients need;
- 2) Serve as a referral source for primary care providers;
- 3) Provide care coordination in collaboration with primary care providers and other specialists;
- 4) Educate local clinicians; and
- 5) Actively conduct surveillance in conjunction with state and federal agencies to monitor trends in complications and health care utilization.

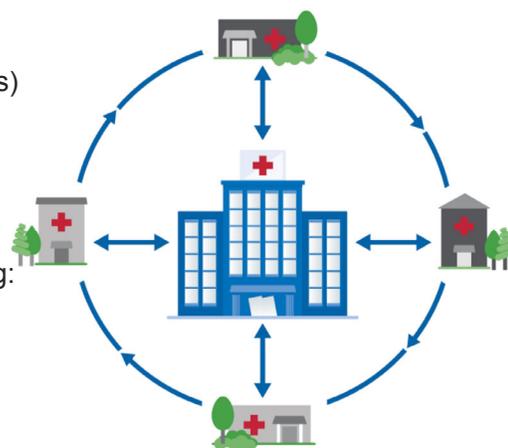


Figure 1. Hub and spoke model for healthcare delivery

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| Strategy 3.3.2 | Provide training and support to adolescents and young adults to increase readiness for transition from pediatric to adult care. |
| Goal 4: POLICY | Ensure Sustainability of California Sickle Cell Action Plan |
| Strategy 4.1 | Identify a mechanism for monitoring policy in California to protect the interests of individuals with SCD and SCT. |
| Strategy 4.2 | Continue collaboration among CA stakeholders. All stakeholders speak using shared messages by creating, implementing, and sharing the CA-SCAP with key partners, decision makers, and the media. |
| Strategy 4.3 | Increase capacity for coordinated stakeholder advocacy regarding sickle cell, especially in the key areas of: <ol style="list-style-type: none"> 1) Availability of trait testing and follow up; 2) Better transition to adult care for adolescents and young adults with SCD; 3) Protecting student civil rights; and 4) The effect of opioid legislation on SCD pain management. 5) Increase capacity for self-advocacy and empowerment among people living with SCD and their families and advocates. |

Conclusion

People living with sickle cell are dying early because of the lack of state infrastructure to address this complex disease.

Taking action will reduce racial and health inequities. It will:

- Help improve the quality of life, productivity and longevity of Californians living with SCD;
- Help people with SCT make informed reproductive decisions; and
- Enhance resource utilization by shifting care from hospitalizations to prevention of complications.

Given that the majority of people with SCD are Medi-Cal beneficiaries, and many expensive sickle cell hospitalizations are avoidable, improving access to comprehensive outpatient sickle cell disease care, particularly for adults, is cost effective for the state. Most importantly, the disparities in health and health care that have pervasively affected individuals with SCD ultimately limits the broader population of Californians in obtaining the highest quality of healthcare, quality of life and the best health outcomes.

Appendix

People Living with SCD in California, 2014-2016

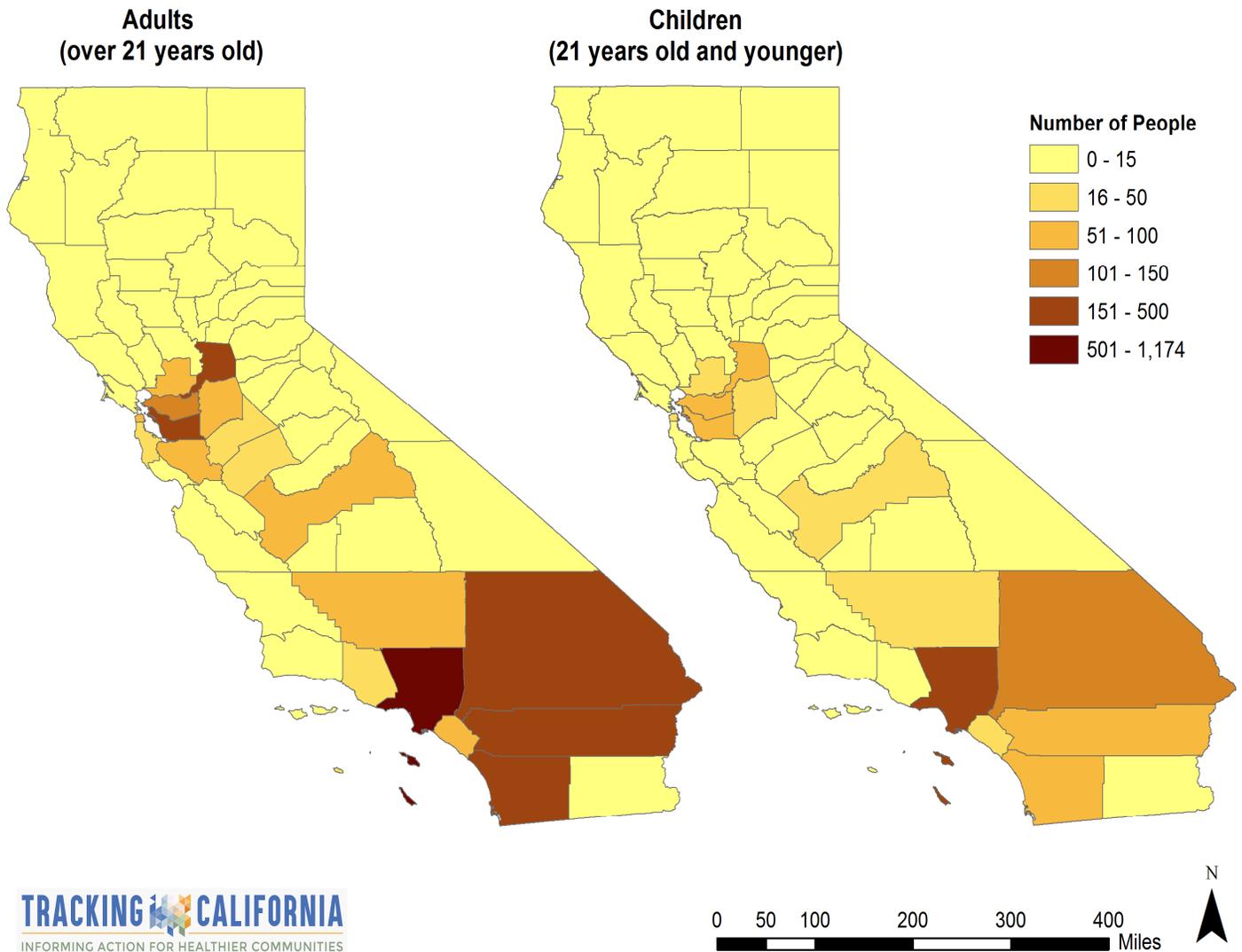


Figure 3. Pediatric and adult populations across the state affected with sickle cell disease

Figure 3 shows the distribution of individuals with SCD in the large metropolitan areas of the San Francisco Bay Area, Sacramento, the Central Valley, and throughout Southern California, including Los Angeles, Kern, Orange, San Bernardino, Riverside and San Diego. At the same time, there are low numbers of adequately trained providers to treat people with SCD in these areas, particularly in Southern California.

This data are from California's current SCD surveillance system—the only initiative that can robustly monitor trends statewide in complications, mortality, services utilization and costs. However, it lacks guaranteed, long-term funding.

Sickle Cell Basics

Sickle Cell Disease (SCD) is caused by a genetic mutation that affects the formation of hemoglobin, the protein in red blood cells (RBCs) that carries oxygen. RBCs become hard, sticky and shaped like a farmer's sickle. These sickled cells clump together blocking blood and oxygen flow in blood vessels. They break down more rapidly than normal RBCs. This can cause a low blood count (anemia).⁵

The abnormal shape of RBCs in people with SCD contributes to problems throughout the lifespan including infections, stroke, and organ damage. SCD is also associated with premature mortality.

Sickle cell trait (SCT) is different from SCD. Sickle cell trait is not a disease, and people with sickle cell trait cannot develop SCD later in life. People with SCT usually do not have any of the symptoms of SCD, but they can pass the sickle cell gene on to their children. Parents who both have SCT have a 1 in 4 chance of each child having SCD.⁶

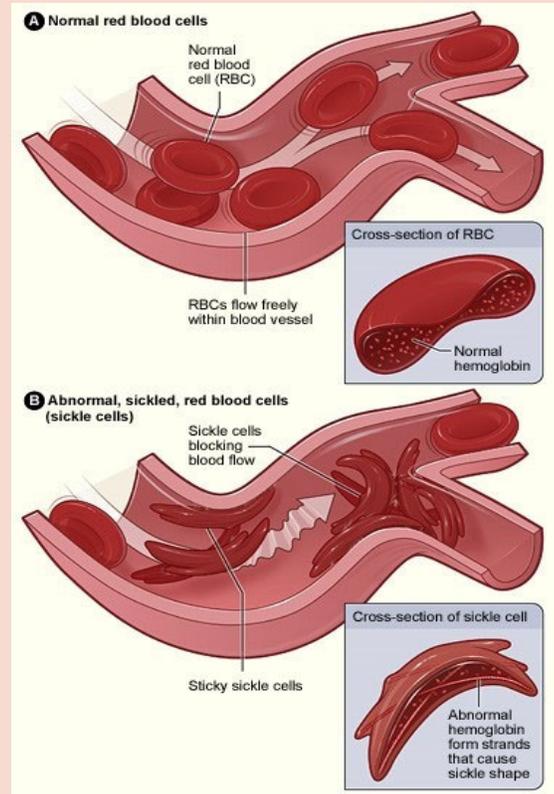


Figure 2. Normal (A) and Sickled (B) Red Blood Cells⁷

Glossary

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| CCS | California Children's Services is a state program that provides diagnostic and treatment services, medical case management, and physical and occupational therapy services to children under age 21 with CCS-eligible medical conditions, one of which is sickle cell disease. Depending on the individual's insurance type, either Medi-Cal, CCS, or CCS Health Families will reimburse healthcare providers for the cost of care. |
| CDHCS | California Department of Health Care Services funds health care services for about 13.5 million Medi-Cal members. DHCS is also responsible for administering the following programs: California Children's Services; Child Health and Disability Prevention program; the Genetically Handicapped Persons Program; the Newborn Hearing Screening Program; the Family Planning, Access, Care, and Treatment program; Program of All-Inclusive Care for the Elderly, Every Woman Counts, and Coordinated Care Management. |
| CDPH | California Department of Public Health implements programs and services in collaboration with local health departments and state, federal and private partners. |
| GHPP | The Genetically Handicapped Persons Program (GHPP) is a health care program for adults with certain genetic diseases, including sickle cell disease. |
| CDPH NBS | Newborn Screening (NBS) is a public health program that screens all babies for many serious but treatable genetic disorders. All babies born in California are required to get screened soon after birth. Newborn screening began in California in 1966 with screening for one disorder, phenylketonuria (PKU). The Program has expanded and now includes 80 different genetic and congenital disorders, including sickle cell disease. The goal of the program is to identify babies with these disorders early, so that treatment can be started right away. |

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