Issue At A Glance:

Disparities in Sickle Cell Disease Funding & Research

Sickle cell disease (SCD) is an inherited blood disorder that predominately impacts individuals from minority backgrounds such as African Americans and Hispanics. Although SCD is the most common inherited blood disorder in the US, this group does not receive adequate federal or private funding. This brief will highlight the funding and research disparity between SCD and cystic fibrosis and provide an overview of two pivotal state and federal sickle cell disease policies.

Introduction

Sickle cell disease (SCD) is an inherited blood disorder caused by a defect in a protein called hemoglobin, which is found in red blood cells and serves to transport oxygen across the body. Due to a structural defect caused by the abnormal hemoglobin, the red blood cells are sticky, rigid, and sickle shaped instead of having a smooth, biconcave disk shape. The sickled cells obstruct small blood vessels and capillaries, disrupting the flow of oxygen to body tissues. As a result, patients experience severe and debilitating pain crises (known as vaso-occlusive episodes) and other complications that affect multiple organ systems.[1,2]

SCD is the most common inherited blood disorder in the United States and affects approximately 100,000 Americans and 3.1 million people globally.[3,4] In the United States, SCD predominately impacts individuals from minority backgrounds, such as African Americans (90%) and Hispanics (10%).[5]

Although SCD is the most common inherited blood disorder in the US, it does not receive adequate federal or private funding. This brief will highlight the funding and research disparity between SCD and cystic fibrosis (CF), a disorder that predominately affects Caucasians, and provide an overview of two pivotal state and federal sickle cell disease policies.

Sickle Cell Disease Statistics

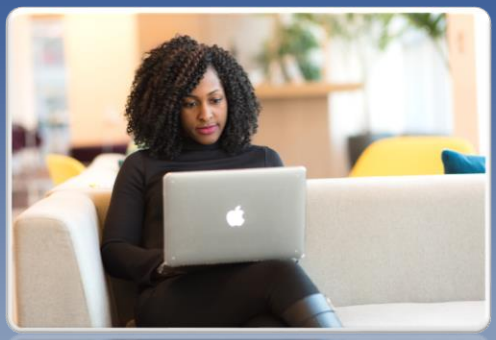
60% of individuals living with sickle cell disease in the United States are adults[6]

90% of individuals living with SCD are Black and suffer significant stigmatization and health disparities[7]

25% of eligible adults with SCD are prescribed hydroxyurea despite its proven efficacy since the early 1990s[8]

Networking California for Sickle Cell Care Initiative

The Networking California for Sickle Cell Care Initiative (NCSCC) is an effort led by the Sickle Cell Disease Foundation (SCDF) and the Center for Inherited Blood Disorders (CIBD). NCSCC is comprised of academic and medical centers across California interested in improving the well-being and health of individuals living with Sickle cell disease (SCD). The goal of NCSCC is to "create sustainable systems of care for SCD patients, particularly the adults, who are among our California's most medically vulnerable population."[9] In addition, NCSCC was established to support workforce expansion for coordinated health services; provide access to specialty care and improve quality of care for adults with sickle cell disease; create a public awareness campaign; conduct surveillance to monitor disease incidence, prevalence, and other metrics; and provide fiscal oversight of the resources.[10]



Funding Disparities Between Sickle Cell Disease & Cystic Fibrosis

Funding for sickle cell disease research, surveillance, prevention, and treatment is disproportionately low. Although sickle cell disease is considered a rare disease in the United States, it ranks low compared to other rare diseases when considering funding and research efforts. A 2020 cross-sectional study highlighted the differences in disease-specific funding between sickle cell disease (SCD) and cystic fibrosis (CF).[8] This study found that federal and private expenditures were greater for individuals living with cystic fibrosis compared with those living with sickle cell disease. They found that the U.S. government-funded research was approximately $2807 per person affected by cystic fibrosis (which predominantly affects white people) compared to $812 per person affected by SCD (which predominantly affects Black people).[8]

The stark difference in disease prevalence makes this funding disparity even more devastating. Sickle cell disease is three times more prevalent than cystic fibrosis, yet between 2008 and 2018, federal government research funding for SCD and CF were similar.[8] To exacerbate matters further, the study found that disease-specific drug developments also favored individuals with cystic fibrosis.[8] At the time of the study, there were two disease-specific drugs for SCD compared with six for CF.

This study highlights the significant funding and drug development disparity between individuals living with SCD and CF. Multiple factors play a role in this disparity and require structural and systemic solutions to adequately address this issue.

Relevant Federal and State Sickle Cell Disease Policies

Background

The funding disparity between SCD and CF is an intricate and complex issue that requires a multifaceted solution. Recently, SCD has gained traction through intensive advocacy and lobbying efforts from devoted advocates, coalitions, community-based organizations, and political leaders, among others. Two federal and state policies are worth noting. These policies go beyond raising awareness by developing collaborative networks and establishing SCD centers of excellence, among other tangible provisions.

Federal Sickle Cell Disease Policy

In 2018, President Trump signed the Sickle Cell Disease and Other Heritable Blood Disorders Research, Surveillance, Prevention, and Treatment Act (S. 2465) into law.[11] S.2465 invested in the development of a national surveillance and prevention program for SCD and other heritable blood disorders, authorizing the award of grants for the following three broad purposes:[11]

1. Collect and maintain data on sickle cell disease health outcomes and carry out various public health activities that include education and training of health professionals at the community, local, and state levels
2. Support local and state laboratories that conduct tests to detect sickle cell disease
3. Evaluate best practices for the prevention and treatment of complications from sickle cell disease

Of note, the Sickle Cell Disease Comprehensive Care Act (H.R. 6216/ S. 3389) was introduced in the Congress on December 14, 2021, and is awaiting the next legislative action.[12]

State Sickle Cell Disease Policy

On June 27, 2019, Governor Gavin Newson signed Assembly Bill (AB) 74, the Budget Act of 2019, into law.[13] AB 74 invests a significant amount of funding toward the advancement of care for individuals living with sickle cell disease in California. AB 74 provides the following provisions:[13]

1. Invest over $14 million to establish a network of sickle cell disease centers throughout the state of California (e.g., Alameda, Fresno, Kern, Los Angeles, Sacramento, San Bernardino, and San Diego) to provide access to specialty care, improve quality of care for adults with SCD, and conduct surveillance, among others
2. Invest $600,000 for the State Department of Public Health to allocate funds to the Public Health Institute (PHI) to support sickle cell disease surveillance and monitoring activities
3. Invest $60,000 to fund administrative activities of the State Department of Public Health in allocating funds to PHI as noted above

In providing these funds, CA took a step forward in improving health disparities faced by those with SCD.

Conclusion

Although federal and state governments are taking steps in the right direction to improve surveillance, prevention, and treatment of sickle cell disease, more work needs to be done. It would behoove SCD advocates to build on this momentum and push for more funding to bolster the provisions offered by these laws. Farooq, et. al. recommends the development of a robust national organization linked with state and local chapters that can pool funds to increase research funding, novel therapeutics, clinical trials, and interconnected comprehensive care centers.[8] This approach is similar to that of the Cystic Fibrosis Foundation, which benefits CF patients nationally. Tackling this complex issue requires collaboration, cooperation, and compassion toward individuals living with SCD.

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Did you know?

The Sickle Cell Disease Center at Loma Linda University Health provides comprehensive services for adults with SCD in California's Inland Empire region, which has the second largest population of individuals with SCD in the state.

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