NICHQ National Institute for Children's Health Quality

Insights

A Physician's Reflections on Racism and Treating Sickle Cell Disease

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HOW SICKLE CELL DISEASE AFFECTS CHILDREN'S HEALTH

Approximately **100,000 people in the U.S. are living with sickle cell disease**, a serious, debilitating, life-shortening, and often fatal condition, which disproportionately affects Black, African-American, and Hispanic-American populations, according to the CDC. In the U.S., SCD is most common among African Americans. However, other racial and ethnic groups are affected, including Latinos and people of Middle Eastern, Indian, Asian, and Mediterranean backgrounds. Sub-Saharan Africa has the greatest burden of disease.

An estimated **2,000 children are born each year in the U.S. with sickle cell disease.** Despite the study of biological pathways for sickle cell disease for more than 100 years and the existence of innovative and effective treatments for this condition, there are still major gaps in accessible, quality care. Additionally, state registries of children born with SCD and adults living with SCD are too few, resulting in inadequate data on the number of people who need care.

The increase in **life expectancy for people living with sickle cell disease rose to 43 years in 2017**, from 29 years in 1979 and 14 years in the mid-1970s. Now, based on life expectancy modeling, babies born with SCD will live an average of more than 50 years. The interventions and treatments promoted by the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program have improved the lives of tens of thousands of people, mitigating some of the harsh effects of the condition, and increasing both length and quality of life.

Scott D. Berns, Chief Executive Officer at NICHQ

There are times in life when new information prompts us to reflect on our past experiences, ask ourselves tough questions, and contemplate how we could have made different choices. Our willingness to experience those moments and how we change after having them is what could make all the difference *for someone else's quality of life*.

I'm drawn back to a time in the 1990s when I had been working in pediatric emergency medicine at a hospital in a large metropolitan area. We saw children who came in with severe pain crisis from sickle cell disease (SCD). While consistent preventative care helps reduce painful side effects and other condition-related issues, acute care is often urgently needed. The most severe complications of SCD can include vaso-occlusive pain crises, bacterial infections, Acute Chest Syndrome, and stroke – all of which are very serious and life-threatening health events, especially for children. I clearly remember children I have cared for – they would arrive writhing in pain, appearing desperate, crying and inconsolable.

Years later, I read about the care disparities for kids of color living with SCD whose families seek treatment for them in the ER. We know that patient experiences of pain are often stigmatized in the healthcare system – and disturbingly more often for Black families – because of underlying concerns about opioid drug-seeking, though research has shown that, commonly, providers actually underestimate pain levels. (Haywood Jr et al., 2013; National Heart Lung and Blood Institute, 2014; Smith et al., 2008)

Reading data like this pushed me to internally replay some of my interactions in the ER years ago. It can be tempting to relive past memories with the best versions of ourselves as the lead, but I now knew the data, and the data didn't care about my best intentions. These days, through the honest, supportive, and often difficult-but-necessary conversations with my colleagues and my own personal commitment, I've accessed frameworks and shared language to develop an equity lens that is continually growing, allowing me to see my past actions from different viewpoints.

Did I treat people without bias?

In recalling the past, all you can do is sit with it – it's uncomfortable. And it's hard to wonder whether or not I always provided equitable care. Did I think the child or caregiver in front of me was drug-seeking (however implicit the bias) and, therefore, did I treat the child differently? I'd like to say "definitely, no way," but there is now a great amount of data that exists on the unconscious dehumanization of Black children, which impacts interactions like these.

And, I wonder, was I as quick to help Black children with SCD in pain as I was a white child who came in with a broken bone?

The answer is data point to "probably not." The data show that I most likely could have done better by those kids. And collectively, we can all do better by the 2,000 children with sickle cell disease born in the U.S. each year.

Our work in sickle cell disease collaboratives spans a decade – half of NICHQ's existence – and was our first project focused on a rare disease. From my lived experience as a dad of a child with a rare disease to my professional experience as a pediatrician to my service as member and chair of the Patient-Centered Outcomes Institute's (PCORI) Advisory Panel on Rare Disease (2018-2021), I've been proud to partner and learn with other healthcare stakeholders and provide crucial input on the direction of healthcare research to improve the delivery of equitable care to people impacted by serious health conditions.

NICHQ PROJECTS RELATED TO SICKLE CELL DISEASE
Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program

Disseminating Results: Missed Sickle Cell Disease Clinic Appointments and the Health Belief Model

READ THE REPORT TO CONGRESS
See data, program information, and recommendations in our recently published Report to Congress, and browse resources for providers in the Model Protocol and Compendium of Tools and Materials.

NICHQ served as the National Coordinating Center for HRSA's Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program from 2017-2021, supporting data collection and compiling information about programs, services, and recommendations from the work of five Regional Coordinating Centers (RCCs) that covered 53 states and U.S. territories.

More than 100 health systems, clinics, and community-based organizations participated in this iteration of the collaborative, leading to more than 25,000 people with SCD being cared for by engaged healthcare providers. Because of expansions in the collaborative service area, all people living with SCD in the U.S. and its territories now have the potential to be reached.

Still, it's a bold goal to reach everyone who lives with SCD. We must refrain from glossing over the reality that even for the children who are receiving care, many are not achieving optimal health. For sickle cell disease, some factors include a reluctance of providers to treat patients with SCD, overt and/implicit bias, and limited expertise to take care of patients with this rare

disease. It could be easy to think the impacts of implicit bias and racism are limited to individual children and adults not receiving adequate care. However, when we see those harmful patterns repeated at scale, it changes the life course of entire generations of families. When viewed through an equity lens, addressing and eliminating disparities in care, funding, and access to treatment for rare diseases like SCD (that disproportionately impact Black and African-American families) is anti-racism work.

Research continues to show that provider bias remains problematic. (Edwards-Maddox, 2021.) Several providers interviewed from this collaborative noted resistance to care for people living with SCD due in part to disease stigma, lack of knowledge or comfort treating the SCD population, institutional and/or provider bias, and overt racism. Whatever the reason, this translates to more suspicion from care providers and lower quality care. Like other systemically marginalized groups of people with special healthcare needs, patients with SCD report refusal to be seen, long wait times in the ER, and feeling like ER physicians and nurses didn't care about them (Linton et al., 2020) – all factors that drive patients to avoid seeking care during pain crises and experience delays in pain medication dispersals. (Jenerette & Brewer, 2010; National Academies of Sciences Engineering and Medicine, 2020; Shapiro et al., 1997).

Considering how important improving care for people with SCD is, the collaborative's three priority areas focused on some of the most pressing issues related to the care of patients with SCD:

1. Increasing the number of providers using **evidence-based treatment** for people living with SCD
2. Enhancing and increasing **provider-to-provider education** and mentoring
3. Improving **access to quality care** through strategies to engage patients and families

Theoretically, if easy-to-administer preventative treatments are available for a rare disease, they would be highly used and widely known about. However, providers in our collaborative noted there are several barriers stopping this from happening. People with SCD may see clinicians and providers in a variety of settings. Not all providers will have up-to-date training about the evidenced-based recommendations for quality SCD care. In addition, there is an undersupply of trained providers. Greater efforts must be made to improve and strengthen the pipeline of knowledgeable providers, such as specifically training advanced practice providers (i.e., nurse practitioners, physician assistants). Areas that the collaborative focused on for evidence-based treatments included use of hydroxyurea (HU), immunizations, and Transcranial Doppler screening.

Regional Coordinating Centers conducted provider-to-provider education to address some of the care knowledge issues. Informal efforts included teaching activities like federally qualified health centers hosting "lunch and learns" to discuss important clinical and psychosocial issues and maintaining connections to resident training programs. More formal efforts included Project ECHO, a globally recognized telementoring program designed to create virtual communities of learners by bringing together healthcare providers and subject matter experts using videoconferencing to host brief lecture presentations and case-based learning.

NEW RESOURCES FOR CARE OF SICKLE CELL DISEASE

New resources include discussion guides for community-based organizations and for providers to use with patients, as well as an infographic for caregivers that prompts reflection on reasons for missed appointments as well as strategies to address them.

High quality care is comprehensive, coordinated, and continues through the lifespan. Often, young people struggle to transition safely from pediatric care to adult care, so that was one area of focus for the collaborative. Coordinated care is important because people with SCD require several levels of care that often involve multiple specialists, several procedures, and serial appointments. NICHQ is currently leading a PCORI project, Disseminating Results: Missed Sickle Cell Disease Clinic Appointments and the Health Belief Model, that investigates the constellation of factors surrounding missed appointments and what providers and community-based organizations are doing to reduce missed appointments.

Currently, the only cure for SCD is bone marrow transplant. However, because of the challenges of finding suitable matches, the risks involved with the procedure, and extensive related costs, in reality, this is rarely an option. Innovative new gene therapy treatments are inspirational but due to their experimental nature and costs, this path currently seems even more inaccessible than standard preventative treatments and therapies. According to the New York Times, the current price tag per patient of up to $2 million for gene therapy treatment is a hard sell for a rare disease, which is compounded by a “legacy of underinvestment” when compared to select diseases that mostly affect white people.

We must vigorously continue to support efforts that discover treatments and the cure for SCD. But we must equally ensure that the screening tests, preventative measures, and treatments are accessible, affordable, and, importantly, equally available to all eligible patients. The search for a cure can serve as our North Star – often this pursuit is what fuels hope, perseverance, and faith for so many. But, as healthcare providers in complex systems riddled with systemic racism, we also need to keep examining what we can do right now to improve the quality of life for the nearly 100,000 people living with this painful disease in the U.S.

For NICHQ’s current and future work, I am motivated by wanting to be a better version of myself in service of others. Wondering whether my own implicit biases impacted my care of patients and families, I realize that I cannot redo past ER experiences. If I could go back, I would slow down to acknowledge my implicit biases and try to set them aside and approach patients from a more informed perspective. But now, I can use my past, present, and future experiences to ensure NICHQ is amplifying important lessons from this multi-year effort reflecting the compassion, care, and commitment of hundreds of dedicated professionals in pursuit of equitable, accessible, and quality healthcare for people living with sickle cell disease.