

When Actions Speak Louder Than Words — Racism and Sickle Cell Disease

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The merciless killing of George Floyd by Minneapolis police officer Derek Chauvin and the more recent shooting of Jacob Blake by Kenosha police officer Rusten Sheskey have catalyzed the most widespread protests in American history, calling not only for an end to police brutality but also for a nation in pain to reflect on the layers of racism that allow such brutality to persist. Medicine is a mirror for the racial injustice in our society; it is a field riddled with racial disparities in everything from research funding to patient care to life expectancy.¹⁻³ There may be no population of patients whose health care and outcomes are more affected by racism than those with sickle cell disease (SCD). Patients with SCD are too often marginalized and dismissed while seeking medical care when their bodies hurt and they cannot breathe. As medical leaders around the United States issue statements denouncing racial injustice and calling for us to “dismantle racism at every level,”⁴ we must ensure that these pledges translate into durable improvements for patients with SCD.

SCD is a life-threatening, inherited blood disorder, affecting more than 100,000 Americans.⁵ Painful vaso-occlusive crises, the hallmark of SCD, result in substantial suffering and lead to associated stigma. Without adequate treatment, SCD affects all organs and is associated with decreased quality of life and a shortened life span. Among the dozens of conditions that are screened for in state newborn-screening pro-

grams, SCD is the most commonly detected condition, regardless of ethnicity. It is thus important to recognize SCD as a common and important medical condition among Americans, and not “just Black Americans.”

The causative mutation in SCD primarily arose on the African continent because of the protective effect of the carrier state against malaria, so most patients with SCD have a shared African ancestry. Although SCD is a global disorder affecting people of all races, in the United States, as a direct result of the transatlantic slave trade, nearly all patients with SCD are Black. This fact would be mere medical trivia if we did not live in such a highly racialized society. Unfortunately, the social construct of race in America requires the majority of patients with SCD not only to face the consequences of a serious health condition, but also to navigate a society in which the color of their skin is often an unfair disadvantage.

“Structural racism” refers to the system of discriminatory policies that assign privilege and power on the basis of race. For example, the legacy of redlining, a policy that allowed banks to deny mortgages to Black Americans who wanted to purchase homes in White-designated neighborhoods, persists today in the form of high rates of housing insecurity among Black Americans, including those with SCD. These racist policies have led to enduring cycles of poverty that result in food insecurity, fewer employment opportunities, and incon-

sistent health insurance coverage, all of which compound the challenges faced by people living with SCD.

Although SCD was first described more than 100 years ago, the development of disease-modifying therapies has stagnated because of inadequate research funding, attributable at least in part to structural racism. Cystic fibrosis is comparable to SCD as an inherited, progressive, life-threatening disease associated with decreased quality of life and shortened life span, but it primarily affects White Americans. Cystic fibrosis affects one third fewer Americans than SCD but receives 7 to 11 times the research funding per patient, which results in disparate rates of development of medications: currently, the Food and Drug Administration has approved 4 medications for SCD and 15 for cystic fibrosis.³ We need to analyze the role of race and racism in the persistence of such discordant resource allocation. Federal funders, lawmakers, advocates, and health care leaders need to work together to reverse the impact of years of structural racism on SCD funding, research, and policy decisions.

In addition to the substantial barriers created by structural racism, the access to and delivery of high-quality health care for patients with SCD is also disrupted by interpersonal racism. Too often patients with SCD simultaneously combat unbearable pain and racist attitudes expressed by health care workers in our hospitals.¹ Despite inexorable pain, patients report getting dressed nicely be-

fore presenting to the emergency department in an attempt to avoid judgment and receive better care. The term “sickler” — a word laced with racist overtones — is often used ignorantly to describe and depersonalize patients with SCD. Deplorably, especially in the midst of the opioid crisis, patients with SCD are often described as drug seekers and accused of feigning their pain, which results in inadequate treatment and more suffering. Because of the challenges in receiving adequate care and the stress associated with perceived racial stigma, many patients choose to avoid care altogether, further increasing the risk of life-threatening complications.¹ It is unacceptable for the medical community to tolerate this racist mistreatment of patients with SCD in our emergency departments and hospitals.

Our hospitals and clinics must be safe spaces for patients with SCD. Racism is deeply ingrained in our society and culture. It informs unconscious judgments and implicit biases, even in well-intentioned clinicians, that can negatively affect the evaluation and care of patients and can be exacerbated in stress-filled emergency departments, hurried clinics, and busy wards.² All health care providers must be open-minded and self-aware as they care for patients with SCD, recognizing their own implicit biases and making a conscious effort to treat patients equitably, regardless of race. They should be required to regularly examine their own racial biases in a supportive environment that helps reduce the impact of these biases on their behavior. Outpatient providers who develop long-term relationships with patients with SCD must be willing to have open, honest, and explicit conversations

Proposed Changes to Reduce the Impact of Racism on Patients with Sickle Cell Disease (SCD) in the United States.

Reduce the impact of structural racism on patients with SCD.

Implement universal screening for social determinants of health in patients with SCD, using connections to available community and governmental resources.
Reintroduce federal funding for comprehensive sickle cell disease centers.
Analyze the effect of race and racism on federal funding for disease research.
Provide psychosocial support for patients with SCD, including social workers, patient navigators, and psychologists.

Dismantle institutional racism with a focus on SCD.

Develop formal, hospital-based reporting systems similar to those for safety events and quality improvement to document and respond to racist behavior.
Include patients with SCD or their advocates on antiracism task forces.
Institute SCD-specific pain-management protocols to reduce the time to opiate administration and to improve health outcomes.
Empower patients with SCD to safely report concerns about racism or inequity.

Address interpersonal racism with patients and colleagues.

Speak explicitly about race within and across medical teams, with a focus on experiences of patients with SCD.
Develop partnerships with patients and recognize their ability to educate providers about the impact of race and racism on their health care experiences.
Implement mandatory annual racial implicit bias training for all clinicians in a supportive environment.
Practice mindfulness and self-reflection in the care of patients with SCD, recognizing that everyone has biases.
Stop using the word “sickler,” and educate colleagues who use it.
Create safe spaces for all health care workers to discuss race and racism and to report events when they happen.

about race and the impact of racism on patients' lives. Finally, inequitable treatment of patients based on race should be reported and addressed as urgently as are other hospital safety events.

United action against racism's impact on the health of patients with SCD is long overdue. The most recent senseless murders of Black Americans and the subsequent wave of pledges to dismantle racism should be our call to action. Change begins with a conversation, but words are not sufficient; we must take action to make enduring improvements in the care of patients with SCD. In the box, we offer a framework and suggestions for initiating change. The current generation of patients with SCD should be allowed to lead full and healthy lives without fear or distrust of the health care institutions whose purpose should be to protect and serve.

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