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The State of Sickle Cell Disease Care in the United States: How Can Emergency Medicine Contribute?



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The Office of Minority Health of the US Department of Health and Human Services (HHS) is honored to sponsor this sickle cell disease (SCD) special supplement in *Annals of Emergency Medicine*. Fittingly, the issue appears in September 2020 during Sickle Cell Awareness Month, an annual observance designated by Congress and widely supported at the national, state, and local levels. The goal of this supplement is to raise awareness about SCD and increase the knowledge of emergency physicians to improve quality of care. SCD is the most common inherited blood disorder in the United States, affecting approximately 100,000 US residents.^{1,2} An estimated 25 million people live with SCD worldwide.

Individuals living with SCD frequently seek care in the emergency department (ED).³⁻⁵ In 1 study, nearly half of the 4,636 identified individuals living with SCD in California had 1 or more ED visits in any given year during 2005 to 2014.³ Complications from SCD, such as pain crises, acute chest syndrome, sepsis, and strokes, often occur acutely, requiring rapid evaluation and treatment. Furthermore, there is a national shortage of SCD-trained hematologists⁶⁻⁸ and primary care providers for adults in the United States, resulting in poor implementation and coordination of chronic care recommendations. This cycle contributes to decreased access and quality of care for individuals living with SCD, particularly adults, and often necessitates the use of EDs as safety-net facilities.

Because SCD is a relatively uncommon condition, most emergency physicians do not treat large numbers of patients with SCD and may not be confident with managing all aspects of the disease. Additionally, as many patients and advocacy organizations have long understood, SCD care—including pain management, prevention, care coordination, and attention to social needs—has not been prioritized. These gaps in care and health disparities have been further exacerbated by the ongoing coronavirus disease 2019 pandemic.

Another persistent challenge for patients with SCD is accessing opioids for acute pain management in the midst of the current national opioid crisis. Restrictive opioid-prescribing practices, an important tool for reducing population-level risk for opioid misuse, have been associated with unintended consequences for patients with SCD and others who rely on ready access to opioids for appropriate care. To raise awareness of the potential harm caused by blanket prescribing policies, the HHS Pain Management Best Practices Inter-Agency Task Force's [final report](#) called for “a balanced, individualized, patient-centered approach” in managing acute and chronic pain and, as an example, outlined 10 SCD-specific recommendations. Similarly, the Centers for Medicare & Medicaid Services [proposed](#) in February 2020 that Medicare beneficiaries with SCD be exempt from drug management programs for opioids and other frequently abused drugs, beginning with plan year 2021.

With this supplement, we aim to highlight the critical role of EDs as frontline health care facilities for SCD care and to identify potential areas for improvement that can be achieved through increased collaboration between emergency physicians, hematologists, and SCD patients and families. The 13 articles in this supplement address broad topics relevant to SCD care, including common clinical presentations in ED settings, as well as emerging diagnostic modalities and barriers to care. Four articles discuss the patient and caregiver experience and psychosocial factors, such as patient stigma and implicit bias.

FEDERAL SCD EFFORTS

HHS has prioritized SCD because of its importance to many US families, but also as an exemplar to organize and implement quality-of-care initiatives for a multitude of relatively rare diseases. Early in 2018, we established an interagency HHS SCD work group to coordinate activities across existing federal programs, reduce duplication, and develop bold, transformative initiatives. The work group is

chaired by the Office of Minority Health and currently comprises 60 members from 12 HHS agencies. The overarching goal of the work group is to increase the life expectancy of individuals living with SCD in the United States by 10 years in 10 years (the 10 × 10 Initiative). President Trump emphasized the importance of SCD by issuing presidential messages in 2018, 2019, and a proclamation in 2020, the first such acknowledgements since 1983.

The SCD Training and Mentoring Program for Primary Care Providers

One new approach to improve patient care is the [SCD Training and Mentoring Program \(STAMP\)](#) for primary care providers using the Project Extension for Community Healthcare Outcomes (Project ECHO) telehealth platform. The target audience for STAMP includes emergency physicians who often serve as de facto primary care providers for patients with SCD. Launched in January 2020, STAMP is a collaboration of the Office of Minority Health and the Health Resources and Services Administration. This 12-part series, taught by hematologists using a case-based approach, covers basic topics of SCD care, such as multimodal pain management approaches, women's health and SCD, and evaluation and management of headache. Participation in STAMP has exceeded expectations, with an average of almost 50 attendees per session, including several emergency physicians. The initial STAMP recruitment strategy focused on national health care associations, including the American College of Emergency Physicians (ACEP), providers from medium-sized cities where needed SCD specialists are lacking, select health care facility types (eg, federally qualified health centers), and SCD advocacy organizations. Through STAMP, a vibrant community of like-minded specialists and primary care/emergency providers is being cultivated, which can potentially seed additional collaborative opportunities to improve SCD care.

Powerful Storytelling of the ED Experience From the Patient Perspective

In February 2020, the HHS Office of the Chief Technology Officer launched the Health+ Sickle Cell Disease Accelerator in partnership with our office. Health+ focuses on specific, high-influence health issues, with the goal of fostering technology solutions and creative problem solving. As an initial step to achieve this goal, the team interviewed more than 25 individuals living with SCD to identify common challenges hindering access to quality care. Two specific topics, ED care and transition from pediatrics to adult care, were developed into [journey maps](#),

which are visually appealing narratives combining data, patient quotes and stories, and graphics. The journey maps enable others to understand more fully the patients' perspective as they navigate the health care system. Both journey maps were used to inform the development of a multistakeholder roundtable and to set the stage for a "Healthathon," launched this month, in which application developers, other technology experts, and SCD stakeholders will meet to develop crowd-sourced solutions addressing some of these challenges.

Model to Increase Hydroxyurea Adherence Using Financial Incentives for Providers

In July 2020, the Office of Minority Health issued a new funding opportunity (\$2.5 million annually for up to 3 years) for state Medicaid offices and partners to develop incentive payment models encouraging providers to increase hydroxyurea use for children living with SCD. Hydroxyurea is a low-cost, long-term medication that has been shown to decrease acute pain crises and episodes of acute chest syndrome, thereby decreasing high rates of ED utilization in this population. The requirements for the grant specify that hydroxyurea performance measures and incentive payment systems be addressed for all major provider types who care for children with SCD, including emergency physicians. The Office of Minority Health developed the grant program based on the premise that all health care encounters are opportunities to educate patients and ensure that evidence-based practice guidelines are being followed. As one example involving EDs, Children's National Hospital in Washington, DC, developed the Quick-Start Hydroxyurea Initiation Project, in which patients with sickle cell anemia, a severe and common form of SCD, who were not receiving hydroxyurea were identified by ED record review and referred for an intensive educational session. Two years after the implementation of the project, the proportion of eligible patients receiving hydroxyurea increased from 56% to 80%.⁹ We know it is essential for emergency physicians to be part of discussions to explore and help inform the development of novel, value-based payment systems. If the payment models are successful, they can be replicated by other state Medicaid offices not only for hydroxyurea but also for other evidence-based practices, both for children and adults living with SCD.

Genetic Therapies, Expanded Data Collection, New Treatments, and a National Blueprint

Hope and optimism for the future of people living with SCD have never been greater than they are now. In

September 2018, the National Heart, Lung, and Blood Institute of the National Institutes of Health launched the [Cure Sickle Cell Initiative](#) to accelerate development of the most promising genetic therapies for SCD so they can be safely used in clinical research within 5 to 10 years. The Centers for Medicare & Medicaid Services issued an SCD indicator in June 2019 based on Medicare and Medicaid claims data and released [2 reports](#) describing SCD prevalence in both these beneficiary populations. In September 2019, together with our office, the Centers for Disease Control and Prevention expanded the number of states that are collecting comprehensive, population-level [data on SCD](#), adding 7 (Alabama, Indiana, Michigan, Minnesota, North Carolina, Tennessee, and Virginia) to the original 2 (California and Georgia). In November 2019, the Food and Drug Administration approved 2 new SCD drugs with novel mechanisms of action, crizanlizumab (monoclonal P-selectin inhibitor) and voxelotor (hemoglobin oxygen-affinity modulator), doubling the number of available SCD therapies from 2 to 4. In addition, more than 35 other candidate SCD therapies are in various stages of review in the Food and Drug Administration pipeline. Finally, earlier this month, the National Academies of Sciences, Engineering, and Medicine issued the nation's first [SCD consensus report](#), sponsored by the Office of Minority Health, which outlines a detailed strategic plan and blueprint to guide future SCD interventions, programs, and research activities.

A CALL TO ACTION

As we engage in these exciting new opportunities, it is essential that we continue to heed and listen intently to individuals living with SCD and to their caregivers. In ED settings, such clear and open communication is crucial for establishing trust, the cornerstone of the patient-physician relationship, which not only affects the quality and effectiveness of care but also can counter previous experiences and assumptions.

We ask the emergency medicine community to partner with hematologists, primary care providers, federal agencies, SCD advocacy organizations, and other stakeholders in using a multidisciplinary, team-based approach to identify solutions that are data driven, patient centered, and compassionate. We encourage wide adoption of warm handoffs between emergency physicians and hematologists, improved awareness and implementation of evidence-based treatment guidelines, and comprehensive coordination of care and supportive services in

communities. It is our hope that this supplement will lay a solid foundation for increasing awareness, identifying key issues, and developing progressive, sustainable collaborations that will help attain the goal of a longer quality lifespan for people living with SCD.

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