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Improving the Quality of Care for Adolescents and Adults With Sickle Cell Disease—It's a Long Road

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The World Health Organization defines quality of care as "the extent to which health care services provided to individuals and patient populations improve desired health outcomes." To achieve this quality, the health care must be safe, effective, timely, efficient, equitable, and centered on people.[1] Kanter et al[2] surveyed 440 adolescents and adults with sickle cell disease (SCD) through 7 comprehensive programs widely recognized as leaders in the care of people with SCD. The results of the survey suggested significant gaps and dissatisfaction with care, especially in the emergency department.

Patient experience has a strong association with the perception of quality of care. Both pediatric and adult SCD populations reported dissatisfaction with care for reasons such as perceived racial discrimination, difficulties with access, and lack of clinician confidence and knowledge in managing SCD complications, especially pain. In the survey described, participants with severe pain or 4 or more pain episodes in the previous 6 months were much less likely to be satisfied with their scheduled nonacute appointments.[2]

In addition, patients with SCD that report experiences of discrimination are 53% more likely to report nonadherence to physician treatment recommendations.[3] Mistrust in health care professionals has been shown to be a mediator of the discrimination-nonadherence relationship. Nonadherence to medical treatment has been associated with increased mortality. These negative experiences and dissatisfaction contribute to treatment nonadherence and avoidance of the emergency department by people with SCD when it is the most appropriate setting. This can lead to adverse outcomes due to delays in care. In addition, the perceived lack of empathy from acute care clinicians described by Kanter et al[2] may be associated with perceived discrimination. People with SCD, frequently present with severe pain without objective findings. This issue coupled with tremendous concern about the risk of opiates and addiction contributes to overt bias, which some patients may report as a lack of empathy on the Adult Sickle Cell Quality of Life Measurement Information System Quality of Care measure. These negative encounters damage trust and create obstacles in therapeutic relationships between patients with SCD and their current and future health care professionals.

Pediatric patients with SCD also report dissatisfaction and perceived discrimination from their clinicians. A study that surveyed parents of hospitalized children on their satisfaction and beliefs about how race influenced how they were treated showed that minority families have a higher dissatisfaction (21% vs 13%).[4] Parents of patients with SCD are 15% more likely to be dissatisfied with their care compared with parents of children admitted for asthma or a general pediatric medical problem. Among African American families, a greater proportion of parents of children with SCD believe that they are treated differently because of race (46% vs 3%-8%; $P < .01$).[4]

Quality of care was assessed in an adult population in the Pain in Sickle Cell Epidemiology Study using the Patient Satisfaction Questionnaire-18.[5] Adults with SCD who received the majority of their care at specialized centers have significantly higher satisfaction scores compared with patients who received most of their care at nonspecialized centers. The difference in satisfaction is associated more with the technical quality of the care rather than with parameters measuring the doctor-patient relationship. That study included a population predominantly in the state of Virginia, in the Southeast United States. More than 300 participants in the Sickle Cell Disease Implementation Consortium are

also being treated in specialized centers in the Southeast US; therefore, findings in both studies may reflect a regional experience. It is unclear whether these findings are generalizable to all regions of the US.

Clinician comfort with caring for patients with SCD is a barrier to improving quality of care specifically in the ambulatory setting. A survey of primary care physicians (PCP) at Johns Hopkins Community Physicians showed that PCPs are not comfortable caring for individuals with SCD.[6] They are most uncomfortable when it comes to managing SCD-specific issues, such as prescribing hydroxyurea and managing pain crises. Prior experience caring for patients with SCD and knowledge from residency are associated with increased confidence. In a focus group, patients with SCD in the United Kingdom reported receiving poor quality of care from their PCPs primarily due to a perceived lack of confidence and knowledge about SCD-specific disease management, a lack of communication, and difficulties with accessibility.[7] Patients still felt that their PCPs had an important role in their general health management.

Several strategies have been proposed to improve quality of care for patients with SCD. System approaches include comprehensive sickle cell centers, hub and spoke models, satellite clinics, and telemedicine. Comprehensive sickle cell centers have shown improved quality of life, reduced health care cost, and reduced health care utilization rates among patients with SCD.[5] This multidisciplinary approach has been shown to improve care in patients with other chronic diseases as well. There are many challenges to the comprehensive center model because it requires substantial resources and highly trained staff and clinicians invested in caring for individuals with SCD. Comprehensive centers can serve as a hub for spokes at community hospitals, satellite clinics, or telemedicine and telementoring programs to expand their reach into rural areas and regions with a lower prevalence of SCD. Legislation to increase funding to improve care for this population has historically had a substantial change in the quality of care and has been shown to be cost-saving in the long term. In addition, there is a large disparity in federal and foundation research funding for SCD compared with cystic fibrosis, a disease substantially less common than SCD in the US and a population with substantially higher average socioeconomic status.[8] This disparity may be associated with less research productivity and novel drug development for SCD. Eliminating this funding disparity may improve the ability to provide more equitable care and thus facilitate a higher quality of care.

The data on SCD self-efficacy from the survey by Kanter et al[2] identified associations with lower self-efficacy and increased pain, emergency department visits, and hospitalizations for pain. Low SCD self-efficacy was associated with older age, disabled employment status, and lower annual income. This finding supports a potential role for patient-level interventions to overcome social determinants of health that may reduce SCD self-efficacy.

How can we reach our destination of high-quality care for all people with SCD in the US? We need to invest substantial resources to expand comprehensive sickle cell centers and to improve SCD self-efficacy. Our efforts need to go down the road into smaller communities and rural areas and overcome the negative influence of social determinants of health that can be barriers to high-quality care.

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