Moving the needle: using quality improvement to address gaps in sickle cell care

Seethal A Jacob,[1,2] Jennifer C Yui [3]

[1]Section of Hematology/Oncology, Riley Hospital for Children, Indianapolis, Indiana, USA
[2]Children's Health Services Research, Indiana University School of Medicine, Indianapolis, Indiana, USA
[3]Division of Hematology, Department of Medicine, Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

Correspondence to
Dr Jennifer C Yui, Division of Hematology, Department of Medicine, Johns Hopkins University School of Medicine, Baltimore, MD 21205, USA; jyui1@jhmi.edu

Accepted 12 July 2023
Published Online First 27 July 2023

► http://dx.doi.org/10.1136/bmjqs-2022-015209

To cite: Jacob SA, Yui JC. *BMJ Qual Saf* 2023;**32**:570–572.

In 2008, the Sickle Cell Society of the UK published clinical standards for the care of adults with sickle cell disease (SCD).[1] Over the subsequent decade, nations including France, Nigeria and the USA also published national guidelines for the management of SCD across the lifespan. However, studies have repeatedly demonstrated that patients are not consistently receiving this evidence-based care, and that up to a third of providers in the USA are unaware of the guidelines.[2]

The barriers to appropriate care of individuals with SCD are complex, given the wide range of manifestations of the disease,[1] the scarcity of haematology and primary care providers and multidisciplinary team members,[2] and complex sociopolitical factors including medical racism and the significant bias that individuals with SCD experience in healthcare settings and the community.[3] In sub-Saharan Africa and India, where the vast majority of individuals with SCD are born and childhood mortality remains high, the barriers are even higher given the limited resources and infrastructure for care.[4] The consequence of this host of factors is poor uptake of and adherence to preventative measures such as pneumococcal vaccinations and transcranial Doppler stroke screening, as well as limited and varied prescribing of established disease-modifying therapies such as hydroxyurea. As a result, focused efforts on quality improvement (QI) initiatives to increase utilisation of such evidence-based screening tests and treatments are needed to enhance access and quality of care delivered to this population.

The rigorous application of QI methodologies provides a path to improving care for individuals with SCD. Effective QI includes engagement of a multidisciplinary team of stakeholders, an outcome-focused approach that requires an extensive evaluation of the current processes and baseline to develop specific aims, incorporation of theories of change to guide the interventions chosen and continuous improvement cycles to best achieve better outcomes within the local environment. Emergency department care in SCD, for example, has been substandard in its provision of high-quality pain management for individuals with SCD, with prolonged wait times, inadequate dosing of analgesia and significant bias in care delivery.[3] As a result, there have been multiple QI initiatives in the emergency department that have focused on reducing the time from an individual with SCD presenting to the emergency department to receiving their first dose of analgesia.[5 6] Similarly, QI methodology has been employed to reduce rates of acute chest syndrome[7] and increase stroke screening.[8] However, while much of the improvement work in SCD employs these principles, teams may not adhere rigorously to QI methodologies, including repeated and iterative Plan-Do-Study-Act cycles, which may lead to less robust results.[9]

In their study published this issue, Alvarez and colleagues[10] completed a number of Plan-Do-Study-Act cycles with various provider and patient interventions that led to significant increase in hydroxyurea prescriptions for children with SCD. In their background work, they identified factors related to provider processes, patient choices and patient environment that were likely to influence hydroxyurea prescription rates. Provider factors at baseline were consistent with those described in previous work, with barriers to prescription uptake including lack of provider awareness of the benefits of hydroxyurea, provider concerns about

side effects and carcinogenic potential of hydroxyurea and provider presumptions of patient adherence.[2 11 12] Educational materials, information on motivational interviewing techniques and electronic medical record reminders of patient eligibility for hydroxyurea were all employed to address provider factors.

To understand the health behaviours around hydroxyurea in their patients, Alvarez and colleagues then employed the health belief model. The health belief model defines six constructs that are hypothesised to predict why individuals engage in health behaviours: perceived susceptibility, perceived severity, perceived benefits of action, perceived barriers of action, cues to action and self-efficacy. The health belief model has been used to promote the adherence to treatments and health behaviours including cancer screenings.[13] The study team surveyed patients and parents on barriers, and found patient concerns consistent with prior studies, and in line with the domains of the health belief model, including perceived lack of severity of their own SCD, lack of perceived benefit, fear of side effects or carcinogenic potential and aversion to taking medications.[11 14] By using the health belief model to better understand their patients' behaviours around hydroxyurea use, the investigators were able to tailor educational interventions and their motivational interviewing to the individual patient's concerns and barriers, and achieved sustained change in hydroxyurea prescription rates.

Integrating these QI methodologies into clinical practice nationwide and worldwide involves incorporating the collection and assessment of quality metrics into the management of sickle cell programmes and use of these data to drive intervention cycles. Raphael and colleagues proposed a health services research agenda to improve health outcomes and survival for paediatric patients with SCD; chief among these strategies were health systems measuring key quality indicators and providers monitoring their adherence to SCD guidelines.[15] However, across much of the world, healthcare governing bodies and stakeholders have not tracked clinically important quality metrics for individuals with SCD, which limits the broad adoption by healthcare systems to meet care standards. One major factor in the ability to develop such quality indicators is limitations in the available evidence and the dearth of randomised clinical trials on the management of individuals with SCD. In the USA, this is compounded by the limited federal and foundational funding for SCD.[16] Despite existing consensus guidelines and evidence-based recommendations that should be employed across SCD centres and accessible to all individuals living with SCD, the ultimate effect is suboptimal quality of care of individuals with SCD and worsening disparities.

Fortunately, efforts are being undertaken worldwide to improve the care of individuals with SCD. In sub-Saharan Africa, the Sickle Pan-African Research Consortium, a multinational collaboration, was established to improve the care of individuals with SCD. The Sickle Pan-African Research Consortium is working to introduce newborn screening in sub-Saharan Africa, and has also adapted available clinical guidelines for healthcare facilities ranging from rural settings without medical expertise to referral hospitals with specialist care available.[17] Additionally, within the USA, large collaborative databases such as the GRNDaD registry allow for data collection on patients with SCD nationwide, and provide a platform for QI initiatives carried out across multiple sites simultaneously.[18] These larger databases may also serve as a bridge between local improvement efforts within a specific clinic or healthcare system, and the implementation of best practices designed to increase the uptake of evidence-based interventions more broadly.

Ultimately, the development of standardised guidelines and longitudinal registries is an important step in improving care delivered to individuals with SCD but is insufficient alone. Worldwide, improving the care of individuals with SCD requires training classical haematologists and other specialists with expertise in SCD. Efforts to improve care in low and middle-income countries may also require support from international health agencies to improve the infrastructure to deliver medication and obtain appropriate laboratory testing for disease and drug monitoring.[19] Also in Western nations, addressing implicit bias and racism in medicine is key to improving care of individuals with SCD.[20]

While the barriers to high-quality sickle cell care are multifactorial and require significant effort to address, one aspect of this can be addressed through QI initiatives, helping to close the gap in SCD care through standardising high-quality, evidence-based treatments across clinics that care for individuals with SCD. The work by Alvarez and colleagues is an example of how this can be employed in a rigorous manner across multiple sites to maximise impact and sustain outcomes.

Contributors SAJ and JCY contributed equally to this paper.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Not applicable.

Provenance and peer review Commissioned; internally peer reviewed.

ORCID iD

Jennifer C Yui http://orcid.org/0000-0003-2375-9530

REFERENCES

1 Society TSC. *Standards for the clinical care of adults with sickle cell disease in the UK*. London, UK, 2008.

2 Smeltzer MP, Howell KE, Treadwell M, *et al*. Identifying barriers to evidence-based care for sickle cell disease: results

from the sickle cell disease implementation consortium cross-sectional survey of healthcare providers in the USA. *BMJ Open* 2021;11:e050880.
3 Haywood C, Tanabe P, Naik R, *et al*. The impact of race and disease on sickle cell patient wait times in the emergency department. *Am J Emerg Med* 2013;31:651–6.
4 Thomson AM, McHugh TA, Oron AP, *et al*. Global, regional, and national prevalence and mortality burden of sickle cell disease, 2000-2021: a systematic analysis from the Global Burden of Disease Study 2021. *The Lancet Haematology* June 2023.
5 Treadwell MJ, Bell M, Leibovich SA, *et al*. A quality improvement initiative to improve emergency department care for pediatric patients with sickle cell disease. *J Clin Outcomes Manag* 2014;21:62–70.
6 Kavanagh PL, Sprinz PG, Wolfgang TL, *et al*. Improving the management of Vaso-occlusive episodes in the pediatric emergency department. *Pediatrics* 2015;136:e1016–25.
7 Davis B, Brooks T, Woolery K, *et al*. Prevention of acute chest syndrome by implementing a standardized process to improve incentive spirometry use in hospitalized patients with sickle cell disease. *Blood* 2017;130:132.
8 Kanter J, Phillips S, Schlenz AM, *et al*. Transcranial doppler screening in a current cohort of children with sickle cell anemia: results from the DISPLACE study. *J Pediatr Hematol Oncol* 2021;43:e1062–8.
9 Knudsen SV, Laursen HVB, Johnsen SP, *et al*. Can quality improvement improve the quality of care? A systematic review of reported effects and methodological rigor in plan-do-study-act projects. *BMC Health Serv Res* 2019;19:683.
10 Alvarez OA, Rodriguez-Cortes H, Clay ELJ, *et al*. Successful quality improvement project to increase hydroxyurea prescriptions for children with sickle cell anaemia. *BMJ Qual Saf* 2023;32:608–16.
11 Treadwell MJ, Du L, Bhasin N, *et al*. Barriers to hydroxyurea use from the perspectives of providers, individuals with sickle cell disease, and families: report from a U.S. regional collaborative. *Front Genet* 2022;13:921432.
12 Lanzkron S, Haywood C Jr, Hassell KL, *et al*. Provider barriers to Hydroxyurea use in adults with sickle cell disease: a survey of the sickle cell disease adult provider network. *J Natl Med Assoc* 2008;100:968–73.
13 Lau J, Lim T-Z, Jianlin Wong G, *et al*. The health belief model and colorectal cancer screening in the general population: a systematic review. *Prev Med Rep* 2020;20:101223.
14 Brandow AM, Jirovec DL, Panepinto JA. Hydroxyurea in children with sickle cell disease: practice patterns and barriers to utilization. *Am J Hematol* 2010;85:611–3.
15 Raphael JL, Kavanagh PL, Wang CJ, *et al*. Translating scientific advances to improved outcomes for children with sickle cell disease: a timely opportunity. *Pediatr Blood Cancer* 2011;56:1005–8.
16 Farooq F, Mogayzel PJ, Lanzkron S, *et al*. Comparison of US Federal and foundation funding of research for sickle cell disease and cystic fibrosis and factors associated with research productivity. *JAMA Netw Open* 2020;3:e201737.
17 Paintsil V, Ally M, Isa H, *et al*. Development of multi-level standards of care recommendations for sickle cell disease: experience from Sickleinafrica. *Front Genet* 2022;13:1052179.
18 Lanzkron S, Manwani D, Desai P, *et al*. Grndad: big data and sickle cell disease. *Blood Adv* 2022;6:1088.
19 Zhou AE, Travassos MA. Bringing sickle-cell treatments to children in sub-Saharan Africa. *N Engl J Med* 2022;387:488–91.
20 Power-Hays A, McGann PT. When actions speak louder than words - racism and sickle cell disease. *N Engl J Med* 2020;383:1902–3.