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 standard 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.\(^5\) 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. 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. 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.

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. 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. 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. Additionally, within the USA, large collaborative databases such as the GRN DaD registry allow for data collection on patients with SCD nationwide, and provide a platform for QI initiatives carried out across multiple sites simultaneously.

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. Also in Western nations, addressing implicit bias and racism in medicine is key to improving care of individuals with SCD.

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.

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

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