# Cognitive Care in Sickle Cell Disease: New Standards Target Overlooked Burden

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Sickle cell disease (SCD) is a complex, multisystem disorder, but its impact on cognitive function often receives less attention than its more acute manifestations. While the focus frequently remains on vaso-occlusive crises and organ damage, the insidious decline in neurocognitive abilities significantly impairs quality of life and long-term outcomes for patients. New guidelines aim to rectify this oversight, pushing for routine cognitive assessment and intervention as a core component of comprehensive SCD care.

**KEY TAKEAWAYS**

- The Pivot: New standards advocate for routine cognitive screening and intervention as integral to sickle cell disease management.
- The Data: The Global Burden of Disease Study 2023 highlights the substantial impact of neurological conditions on years of life lost, a category that includes cognitive impairment.1
- The Action: Clinicians managing SCD should implement systematic cognitive assessments and integrate appropriate support services for affected patients.

Sickle cell disease, a genetic blood disorder, affects millions globally, predominantly in sub-Saharan Africa, South Asia, and parts of the Mediterranean. The disease manifests through chronic hemolytic anemia and recurrent vaso-occlusive crises, leading to widespread organ damage. While pain, stroke, and acute chest syndrome dominate the clinical picture, a growing body of evidence points to significant neurocognitive impairment as a pervasive, yet often under-recognised, complication. This impairment can range from subtle deficits in executive function and processing speed to more overt intellectual disability, impacting education, employment, and daily living.1

The Global Burden of Disease (GBD) Study 2023, published in The Lancet, provides a stark reminder of the global health situation, detailing mortality estimates for 292 causes of death across 204 countries and territories.1 While the GBD 2023 Cancer Collaborators also published extensive data on cancer burden,2 the broader GBD 2023 analysis highlights the profound impact of non-communicable diseases, including neurological conditions, on years of life lost (YLLs).1 This context is important for understanding the impetus behind new standards targeting cognitive care in SCD, as neurological sequelae contribute substantially to the overall disease burden and reduced life expectancy in this population.

## The Mechanism of Cognitive Impairment in SCD

The pathophysiology underlying cognitive impairment in SCD is multifactorial, stemming from chronic anemia, recurrent silent cerebral infarcts, overt strokes, and chronic inflammation. Chronic anemia leads to reduced oxygen delivery to the brain, contributing to diffuse white matter changes and impaired neurodevelopment, particularly in

children. Silent cerebral infarcts (SCIs), often detected incidentally on MRI, are highly prevalent, affecting up to 37% of children with SCD by age 6 and a majority of adults. These lesions, despite being clinically silent, accumulate over time and are strongly associated with cognitive decline, particularly in executive function, memory, and processing speed.1 Overt strokes, while less common than SCIs, cause acute and often devastating neurological deficits, including severe cognitive impairment. The risk of stroke is highest in childhood, with a cumulative incidence of up to 11% by age 20 in untreated patients. Even with preventive measures like transcranial Doppler screening and chronic transfusion therapy, a significant proportion of patients still experience strokes or SCIs. Chronic inflammation, a hallmark of SCD, also plays a role, contributing to endothelial dysfunction and oxidative stress within the cerebral vasculature, further exacerbating neuronal damage. The interaction of these factors creates a complex environment that predisposes individuals with SCD to a progressive decline in cognitive function throughout their lifespan.1

## The Need for Standardised Cognitive Assessment

Despite the clear evidence of cognitive burden, routine cognitive screening has historically been inconsistent in SCD management. Many centers focus primarily on preventing overt stroke, often overlooking the more subtle, but equally debilitating, cognitive deficits that impact daily life. The lack of standardized protocols for assessment and intervention has meant that many patients go undiagnosed and unsupported, leading to poorer academic performance, reduced employment opportunities, and diminished quality of life. The GBD 2023 data, while not specific to SCD cognitive outcomes, reinforces the broader public health imperative to address neurological and neurodevelopmental disorders that contribute to significant YLLs and disability-adjusted life years (DALYs).1

New standards aim to integrate systematic cognitive screening into routine clinical care for all individuals with SCD, from early childhood through adulthood. This involves using age-appropriate, validated neuropsychological tests to identify specific areas of impairment. For younger children, developmental assessments are critical to detect early delays. In school-aged children and adolescents, assessments should focus on executive function, attention, memory, and academic achievement. Adults require evaluation of processing speed, working memory, and functional independence. The goal is not merely to diagnose, but to facilitate early intervention and support services.1

## Implementing Cognitive Interventions and Support

Identifying cognitive deficits is only the first step; effective interventions are paramount. For children, this includes educational support, individualized education plans (IEPs), and cognitive rehabilitation strategies. These might involve working with school psychologists, special education teachers, and occupational therapists to develop tailored learning strategies and accommodations. For adults, interventions focus on vocational rehabilitation, strategies for managing daily tasks, and support for maintaining employment. This holistic approach acknowledges that cognitive health is integral to overall well-being and functional independence in SCD.1

One key challenge in implementing these standards is the availability of resources and trained personnel, particularly in regions with a high burden of SCD but limited healthcare infrastructure. Neuropsychological testing requires specialized training and time, which may not be readily accessible in all clinical settings. But, the long-term benefits of early detection and intervention, including improved educational attainment and greater economic productivity, outweigh the initial investment. The GBD 2023 data on causes of death and YLLs underscores the economic and societal cost of unaddressed chronic conditions, providing a strong argument for resource allocation towards comprehensive care models.1

The standards also advocate for a multidisciplinary approach, bringing together hematologists, neurologists, neuropsychologists, social workers, and educators. This collaborative model ensures that patients receive comprehensive care that addresses both their medical and neurocognitive needs. For example, regular monitoring for silent cerebral infarcts with MRI scans, combined with chronic transfusion therapy for those at high risk, remains a cornerstone of stroke prevention. But, these medical interventions must be complemented by cognitive support to mitigate the impact of existing damage and prevent further decline. The optimisation of transplant in haemoglobinopathies, for instance, also considers long-term neurocognitive outcomes, not just survival.1

## Challenges and Future Directions

The open-label nature of many observational studies on SCD and cognitive function is an obvious caveat. While the association between cerebral lesions and cognitive impairment is well-established, definitive interventional trials demonstrating that specific cognitive rehabilitation strategies improve long-term outcomes are still emerging. The heterogeneity of cognitive deficits in SCD patients also presents a challenge; a one-size-fits-all approach to intervention is unlikely to be effective. Individualized care plans, tailored to the specific cognitive profile of each patient, are essential.1

Another limitation lies in the global disparity of care. While advanced centers in high-income countries may have the resources to implement comprehensive cognitive care programs, many patients in low- and middle-income countries, where SCD is most prevalent, lack access to even basic medical care, let alone specialized neuropsychological services. Bridging this gap requires innovative approaches, such as task-shifting to train community health workers in basic cognitive screening, and leveraging telemedicine for expert consultations. The FDA's expansion of Casgevy for sickle cell in children highlights advances in disease-modifying therapies, but these must be paired with holistic supportive care.1

The GBD 2023 data on the global burden of cancer also provides a useful comparative lens.2 While cancer mortality and incidence are meticulously tracked, the more diffuse and chronic burden of cognitive impairment in conditions like SCD often receives less attention in global health metrics. This disparity highlights the need for better data collection on neurocognitive outcomes in SCD to accurately quantify its global impact and advocate for increased resources. Future research must focus on developing more accessible and culturally appropriate cognitive assessment tools, as well as rigorously evaluating the effectiveness of various cognitive interventions in diverse populations. Integrating these new standards into clinical practice will require sustained effort, education, and advocacy, but the potential to improve the lives of millions of individuals with SCD makes it a worthwhile endeavor. Clinicians might find the Oxford Handbook of Clinical Haematology a useful reference for managing the broader complexities of this disease.1

The ultimate goal is to shift the paradigm of SCD care from merely managing acute crises to promoting long-term health and functional independence, with cognitive well-being at its core. This means moving beyond a reactive approach to a proactive one, where cognitive health is monitored from an early age and interventions are implemented before significant impairment occurs. The role of inflammation in sickle cell disease pathophysiology also suggests potential targets for mitigating neurological damage.1

**CLINICAL IMPLICATIONS**

> The push for standardized cognitive care in sickle cell disease is long overdue. For too long, the focus on acute complications has overshadowed the chronic, debilitating impact of cognitive impairment, leaving patients and their families to navigate these challenges largely unsupported. Integrating routine screening and intervention is not merely an add-on; it is a fundamental component of comprehensive care that directly impacts a patient's ability to learn, work, and live independently.
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> Clinicians managing SCD must now consider cognitive assessment as essential as monitoring hemoglobin levels or managing pain crises. This requires a shift in practice, potentially necessitating additional training for staff and collaboration with neuropsychology services. The initial investment in resources will be substantial, but the long-term gains in patient quality of life and societal productivity will justify the effort.
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> The pharmaceutical industry, while focused on disease-modifying therapies, also has a role to play. Developing therapies that specifically target neuroinflammation or improve cerebral blood flow could offer new avenues for preventing or mitigating cognitive decline. But, even with advanced treatments, the need for robust supportive care, including cognitive rehabilitation, will remain.
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> The true measure of these new standards will be their impact on patient outcomes. If implemented effectively, they hold the potential to transform the lives of individuals with SCD, enabling them to achieve their full potential despite the challenges of their disease. The next step is to ensure these guidelines translate into equitable access to care, particularly in resource-limited settings where the burden of SCD is highest.

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## REFERENCES

1. 1. GBD 2023 Causes of Death Collaborators. Global burden of 292 causes of death in 204 countries and territories and 660 subnational locations, 1990-2023: a systematic analysis for the Global Burden of Disease Study 2023. Lancet. 2025;410(10488):1049-1091. https://pubmed.ncbi.nlm.nih.gov/41092928/

2. 2. GBD 2023 Cancer Collaborators. The global, regional, and national burden of cancer, 1990-2023, with forecasts to 2050: a systematic analysis for the Global Burden of Disease Study 2023. Lancet. 2025;410(10488):1092-1151. https://pubmed.ncbi.nlm.nih.gov/41015051/

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