# Sickle Cell Disease in Africa: Navigating Challenges and Embracing Opportunities

Dr. Munube defines what Sickle Cell Disease is, what its clinical manifestations are, and highlights the current treatment.

## Background

Sickle cell disease (SCD) encompasses a group of inherited red blood cell disorders, with sickle cell anemia being one of its manifestations. This autosomal recessive disorder of the □-globin gene leads to clinical manifestations such as hemolytic anemia and recurrent vascular occlusion episodes. The presentation and progression of SCD in patients are diverse, influenced by factors like environment, sickling extent, vascular components, platelets, leukocytes, and plasma proteins.

In Africa, SCD is particularly prevalent in Sub-Saharan Africa (SSA), with some countries reporting a maximum prevalence of 3% (see Figure 1). Despite the United Nations' recognition of sickle-cell anemia as a public health problem in 2008, focused efforts to assess and mitigate the burden of SCD in Africa, where 85% of affected children are born, remain insufficient. The World Health Organization (WHO) proposed a strategy in 2010, acknowledging SCD as a significant cause of child mortality in many African countries. Investments in public health programs, akin to those for HIV or malaria, could include widespread screening, health education, and treatment to prevent or manage SCD complications.

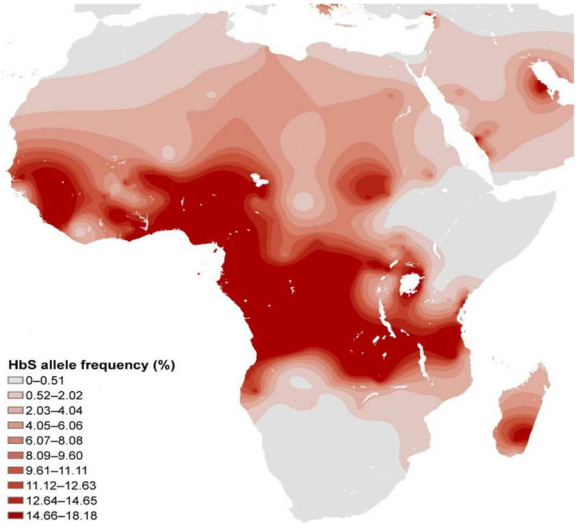


***Figure 1: Map of the distribution of the S gene in Africa ( adapted from Figure 1b in Piel et al.)***

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Mortality rates for SCD children in SSA before the age of 5 range from 50–90%, a stark contrast to improved life expectancy in Western countries. Newborn screening programs, with interventions like penicillin prophylaxis and pneumococcal vaccination, have contributed to this improvement. However, such initiatives are not widely adopted in SSA due to concerns about affordability, operational challenges, and competing priorities, particularly in regions where malaria is endemic. In contrast, life expectancy for SCD patients has improved in Western countries, with most patients living into their 40s and 50s. In those countries, newborn screening programs were introduced incorporating testing newborns for SCD plus interventions (penicillin prophylaxis and pneumococcal vaccination) to prevent bacterial infections among diagnosed cases. Although newborn screening was shown to be cost-effective among African American infants, this approach has not been widely adopted in SSA, where malaria is endemic, and newborn screening programs require an additional component for malaria prevention.

Despite the high SCD burden in SSA, newborn screening and prophylactic intervention programs have not been widely implemented due to concerns of affordability, costs, operational challenges, and competing priorities

## Clinical manifestations

The clinical manifestations of SCD vary from infancy to adulthood. In infants, dactylitis (swelling of hands or feet Figure 2) is common, along with moderate to severe anemia and recurrent chest infections. As individuals age, manifestations such as leg ulcers in adolescents, stroke events starting at age two and peaking at ten, and priapism in teenage years may occur.

## Evidence-Based Interventions

In Africa, clinics commonly provide folic acid, instructions to increase fluid intake, and malaria prophylaxis, although practices vary among countries. Pneumococcal vaccination is part of the national immunization program. Booster doses are subsequently required as a result of the functional asplenia in people living with sickle cell disease. Hydroxyurea, a disease-modifying agent, has shown efficacy in improving the lives of individuals with SCD. While its use is gaining traction, concerns about affordability, costs, operational challenges, and competing priorities still hinder widespread adoption.

Others use alternative anti-malarial medicines for the prevention of malaria. During hospitalization, analgesics, intravenous fluids, antibiotics, and blood transfusion are provided.

*Figure 2: Dactylitis*

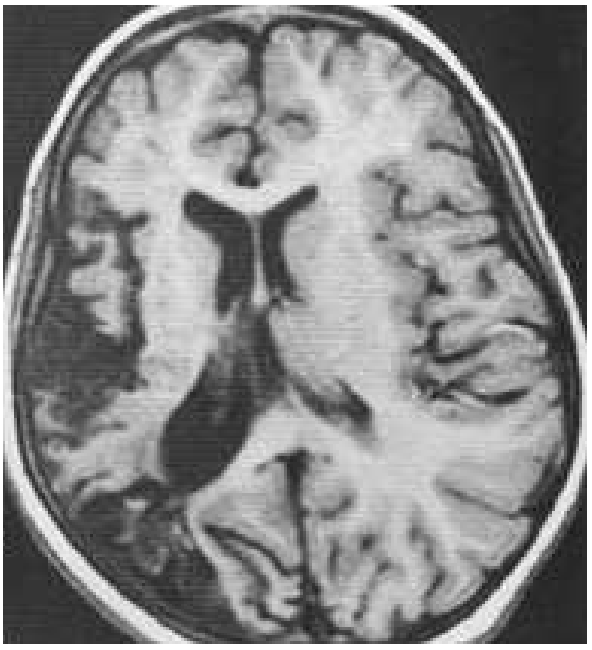

*Figure 3: MRI showing brain Infarcts*

## Hydroxyurea and Its Use in Uganda: A Case Study

Hydroxyurea, a US Food and Drug Administration (FDA) and European Medicines Agency (EMA) approved treatment for SCD, has been included in Uganda's clinical guidelines for regional and national referral hospitals. The guidelines specify its use in severe cases, such as pain crises, strokes, and acute chest syndrome, aiming to reduce the need for blood transfusions and hospitalizations. "Hydroxyurea at a standard dose of 20mg/kg/day should be considered for use in children below 5 years for the management of severe form of sickle cell disease where minimum monitoring conditions and appropriate formulation are available." The severe forms of sickle cell disease have been listed as: pain crises (more than 3 in a year), primary stroke, transfusions (more than 2 in a year), acute chest syndrome, hospitalizations for Sickle cell anemia, and rapid enlargement of the spleen. The use of hydroxyurea will reduce the need for most of the above-stated indications and will have an overall improvement in the lives of people with SCA.

## Lancet Hematology Commission

Following the 2023 launch of the Lancet Hematology Commission on SCD, key recommendations focus on improving health outcomes in Africa and globally. These include standardized epidemiological data collection, expanded screening programs, comprehensive care packages, development of stem cell therapy centers, and healthcare worker training. Progress must be documented through research publications to enable African governments to track improvements in SCD care.

## Conclusion

In conclusion, this article provides an overview of SCD in Sub-Saharan Africa and advocates for early diagnosis, comprehensive care, and treatment in the region. Addressing challenges and embracing opportunities will contribute to better outcomes for individuals living with SCD in Africa.

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