

Àgô Sankofa: an overview of the progression of sickle cell disease in Brazil in the past two decades

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THEMATIC ARTICLE

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Abstract Sickle cell disease (SCD) is an emblematic case of historical health neglect in Brazil and reflects how institutional racism produces health inequalities. This article engaged in a historical journey of this disease, showing the delayed implementation of health policies for people with sickle cell disease, often concealed in Public Power's (in)actions and omissions. The lack of commitment to implement the recommendations of the Brazilian Ministry of Health, such as neonatal screening, and the difficulty in incorporating technologies for health care result from this *modus operandi*. The advances and setbacks in programmatic actions and the constant pressure on several governmental entities have characterized the reported saga in the last twenty years. The present text discusses the policies for people with SCD, appropriating the Sankofa symbol, meaning that building the present is only possible by remembering past mistakes. Thus, we recognize this trajectory and this historical moment in which there is a concrete possibility of moving forward and achieving the longed-for comprehensive care for people with SCD. There is an invitation to glance at a new perspective, one in which hope is the trigger for the movements needed to guarantee the rights of people with SCD.

Key words Anemia, Sickle cell, Black population, Systemic racism, Health policy

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Introduction

Brazil's contribution to generating scientific knowledge about Sickle Cell Disease (SCD) worldwide is relevant, albeit with some distance. It ranks only behind the United States of America, England, and France^{1,2}. From a public policy perspective, the incorporation of the National Policy for the Comprehensive Healthcare of People with Sickle Cell Disease and Other Hemoglobinopathies (PNAIPDF) into the Unified Health System (SUS) through social movements and institutional stakeholders reinforces the leading role and importance of this agenda in Brazil.

If, on the one hand, this prominent role is cause for recognition, on the other, understanding the imbrications of systemic racism in the bowels of global, regional, local, and interpersonal structures hinders the reverberation of this power in the daily lives of people with SCD^{3,4}. It is no coincidence that racism as a system unfolded in different dimensions underpins the theoretical explanatory model for pain, which is SCD's hallmark⁵.

Undoubtedly, SCD is a prototype for understanding how social determination affects the health-illness-care-death process, closely related to the fragmented healthcare model and the low translation of scientific knowledge. Much more than a genetic disease, SCD in Brazil is an emblematic case of how these two cogs – racism and social exclusion – feedback on each other⁶⁻⁸. It illustrates the status quo for experiences, achievements, advances, and setbacks.

The present day still portrays the reality described in 1979: *Some clouds of concern now appear on the horizon for the national sickle cell disease program. There is a flagrant general attention by the Black population and a diluted interest and visibility of the problem of sickle cell anemia caused by political maneuvers to place the program under the legislative umbrella of many other genetic diseases. Moreover, the federal program recently eliminated six reference centers for comprehensive sickle cell disease care and imposed budget cuts on the remaining centers. The victims of this disease, the black population in general, and the researchers and investigators seeking ways to control this disease need assurance from the current national administration that the sickle cell program will not be allowed to die a slow death due to financial attrition, attenuated interest, and willful neglect, leading to the phasing out of another minority project*⁹ (Roland B. Scott, 19789 apud Oransky, 2003, p. 973). Despite the contextual

and historical differences, low interest, budget cuts, political maneuvers, and neglect remain as the Brazilian SCD social history records¹⁰.

SCD is one of the most epidemiologically relevant hereditary hematological diseases, as it is a public health issue in Brazil and worldwide. It affects approximately 30 million people¹¹, with an estimated 300,000 children born alive every year¹². In Brazil, 60,000 to 100,000 people are estimated to have SCD. As a programmatic action, diagnosis was established in 2001 by the national neonatal screening program. Adherence to and coverage of neonatal screening by the states has varied significantly over time. SCD is heterogeneously distributed in the country, and Bahia, the Federal District, and Piauí are the states with the highest number of cases.

Undoubtedly, inaccurate data (under-registration) on SCD can influence the accuracy of its prevalence in the country¹³. Registering all people diagnosed with SCD would provide more reliable information to overcome this inaccuracy. It would allow recording cases diagnosed during prenatal care or blood donations when the diagnosis is established outside of neonatal screening. One indicator refers to the number of children diagnosed with the disease in neonatal screening regarding live births screened¹⁴, which would enhance the possibility of developing other monitoring indicators. The lack of qualified information for health care management and decision-making is a characteristic of the poor implementation of the National Comprehensive Health Policy for the Black Population (PNSIPN)¹⁴ and is a challenge to overcome.

In Brazil, people with SCD life expectancy is approximately 37 years at the median age of death, compared to the general population. In a study on mortality in the country, 3,320 of the 6,553,132 deaths recorded were of individuals with SCD. Among them, the median age at death was 32 years, while it was 69 years in the general population¹⁵.

SCD's relationship with social determination processes is historical and renewed since no policy exists to prioritize this population. In the current situation of economic, political, and health crises, the impoverishment of the Black population is an additional obstacle for households living with a chronic disease like SCD. Whether it is the mothers/grandmothers who stop working to look after the children, the children who drop out of school early because of multiple hospitalizations, or the adults' difficulty in keeping themselves employed amid the periodic crises

caused by the disease, these complex factors hinder social mobility and affect the income of these households.

The recent attempts to dismantle the Unified Health System adversely affect the health of the Black population, who mostly use the public health system. Moreover, the underfunding of research, primarily that focused on socially vulnerable populations, results in a lack of data on the impact of crises on people living with SCD. It is, therefore, clear that past and present are intertwined in the social history of SCD in Brazil.

Given this context, this article aims to show and discuss the persistent reproduction of neglect when it comes to a disease prevalent among the Brazilian Black population, the majority population group in the country. To this end, structural and institutional racism will be analyzed as an explanatory tool for reproducing this neglect. Thus, the fight for the life and health of people with SCD also becomes a fight against racism and racial inequalities in health.

To begin this reflection, we need to ask ourselves: What does the history of policies for people with SCD have to teach us? What does the current Brazilian political situation represent for an agenda neglected for many years? How far have we come? How much further must we go to provide comprehensive care for these people? Is there room for hope? These reflections are in the sessions:

1 – Progress, impasses, and setbacks in the fight for the health of people with sickle cell disease; 2 – Historical and current neglect and the impact of racism on the history of SCD in Brazil e 3 – The current situation of sickle cell disease: concern and hope.

1) Progress, impasses, and setbacks in the fight for the health of people with sickle cell disease

The history of the Comprehensive Care Policy for People with Sickle Cell Disease is linked to the Black population's demand for the right to health. It portrays the history of exclusion endured by this population and the impact of this process on living conditions and health. Thus, the issues of social exclusion, racism, and health inequalities are essential factors in better understanding the social trajectory of SCD in Brazil and the marks left on the bodies of people with the disease.

The construction of this “timeline”, which begins in 1996 and extends to the present day,

allows identifying the main events that have marked the struggle of people with SCD for the right to health. Three significant milestones stand out in this timeframe: inclusion in the National Neonatal Screening Programme (PNTN) and the establishment of the National Federation of Associations of People with Sickle Cell Disease (FENAFAL), both in 2001, and the creation of the National Policy for the Comprehensive Care of People with Sickle Cell Disease and other Haemoglobinopathies (PNAIPDF) in 2005.

The organization of people with SCD and their families into associations and support groups was an important initiative that added to the mobilizations led by the Brazilian Black Movement since the Zumbi dos Palmares March for Citizenship and Life in November 1995, which called for actions to care for the health of the Black population, and participatory management, social control, knowledge production, training and continuing education for health workers to promote equity¹⁷.

President Lula's first term in office was crucial for the Black movement, especially after creating the Secretariat for Policies to Promote Racial Equality (SEPPIR). In 2001, the Third World Conference Against Racism, Racial Discrimination, Xenophobia, and Related Intolerance was held in Durban, South Africa. Brazil's intense participation, especially that of the Black women's movement, enabled progress on the national agenda. Since then, the invisibility of the SCD in public and health policies has been addressed as an emblematic case of neglect and institutional racism.

One of the achievements of this mobilization was the inclusion of SCD as one of the diseases to be screened by neonatal screening. However, although the Ordinance establishing this inclusion was signed in 2001 (Ordinance N° 822 of June 6, 2001), it is only realized in 14 states. The others gradually joined in until 2010. This delay adversely affects the disease's early detection, delaying follow-up onset and impacting the planning of public policies and the very quality of life of people born with the disease. The gains following the inclusion of SCD in Newborn Screening stemmed from intense mobilization by people with SCD and their families.

The National Federation of Associations of People with Sickle Cell Disease (FENAFAL) was established in 2001, with developments in the states that started setting up state associations. In Bahia, the Association of People with SCD in the State of Bahia (ABADFAL) also started its work

this year. Until 2004, the treatment of hemoglobinopathies was one of the National Health Surveillance Agency (ANVISA) activities, which was a hindrance because ANVISA is not responsible for direct healthcare actions.

In 2005, Ministerial Order n° 1.391 established the National Policy for the Comprehensive Care of People with Sickle Cell Disease¹⁸, which was now managed by the Blood and Blood Products Coordination Office (CGSH) in the Secretariat for Specialized Health Care (SAES), which was a significant achievement, as the 2004-2007 Multiannual Budget Plan earmarked resources for the policy for the first time.

Removing the SCD Program from ANVISA and establishing the PNAIPDF Coordination Office was crucial to gaining political backing in the Ministry of Health and beginning to build a care policy in the SUS. During this same period, the term “sickle cell anemia” was replaced by “sickle cell disease”, which more broadly encompasses other hemoglobinopathies, markedly characterized by the predominant presence of the Hb S mutation.

The publication of clinical protocols, ordinances, and resolutions marked a period of intense action by the national management under the coordination of Joice Aragão de Jesus, whose political context was favorable to affirmative action policies in health.

Another Zumbi March was held in Brasilia in 2005, 10 years after the first, in which the Black Movement demanded compliance with the promises in favor of the population with SCD. In 2006, Health Minister Agenor Álvares recognized racism as a health issue during the Second National Seminar on the Health of the Black Population, citing sickle cell disease as an emblematic case.

The ten years since the publication of this decree show that health policies almost always follow a path of comings and goings when they aim to respond to a population's needs. This path is not linear, as there are many battles to break the inertia often imposed by the system. Even achievements already legitimized through ordinances and official documents are under continuous attack, requiring mobilization and pressure from social movements.

The lack of knowledge among health professionals about SCD, both regarding diagnosis and treatment, was so alarming that it resulted in a large number of late diagnoses, a lifetime of hospitalizations, and long years of pilgrimage to health services, often without a diagnosis. However, Brazil's first studies on sickle cell anemia have been conducted since the 1940s.

These studies increased in the 1950s, including findings from population genetics, studies on natural selection, and molecular biology. Thus, an intense production of scientific articles published in journals with wide national circulation and presentations at annual hematology congresses was observed. SCD is incorporated into almost all textbooks as an example of the Mendelian genetic inheritance model, which allows us to reflect on the gap between intellectual production and training in different environments, whether in schools or university health courses.

Giving visibility to the disease has become one of the flagships of the fight, and early diagnosis is a significant measure to reverse SCD's invisibility, with the possibility of having consistent data to support the planning of actions. Incidence data produced by Neonatal Screening can be considered the best data source on the relevance of SCD in Brazil, especially in the Northeast and Southeast, where there is a higher concentration of the Black population. However, some municipalities still need to cover neonatal screening fully, which is even more precarious in small municipalities. Moreover, SCD underreporting is recurrent and stems from health professionals' need for more preparation/interest in completing medical records. The medical records of people with SCD often do not show the corresponding ICD (International Classification of Disease Number) for SCD (D-57) as the underlying disease, which leads to a lack of data or poor quality data on SCD in information systems.

Also in 2011, the Blood and Blood Products Coordination Office launched Technical Note n° 035/2011¹⁹, which guided the inclusion of the Hemoglobin Electrophoresis test in Prenatal Examinations – *Rede Cegonha* (Stork Network), understanding the importance of early diagnosis during prenatal care. This initiative emphasizes the need to examine women's health from a racial perspective. It also proposes that structuring an organized network with primary care as its support and the guarantee of a referral system would have a more significant impact on the morbimortality profile and quality of life for pregnant women with sickle cell disease. We should question the logic adopted here since primary care is primarily the gateway to the health system and is responsible for organizing and coordinating the health actions and services of the territorialized SUS and making referrals to the other care levels.

Through Decree n° 872 of November 6, 2002, the Ministry of Health approved hydroxyurea (HU) use for people with SCD. The drug's progressive use has significantly improved the qual-

ity of life of people with this condition over the last fifteen years, especially among the adult population²⁰. Studies show a reduced mortality of up to 40% for these patients using HU²¹. The drug elevates fetal hemoglobin levels, improving clinical severity and hematological parameters^{21,22}. Steinberger et al.²³ followed people with sickle cell anemia for seventeen and a half years. Mortality declined among those who used HU, and the survival curves showed a significant reduction in the number of deaths with long-term exposure. Approximately 24% of these deaths were due to pulmonary complications, and 87.1% occurred in patients who had never taken HU.

Due to the troubled political scene in 2015, a budget forecast was needed to develop the policy's actions. Even so, the publication of the manual *Sickle Cell Disease: Basic Guidelines for the Care Line* provided states and municipalities with guidelines for including SCD in the comprehensive care network, presenting a care flow whose strategies aimed to overcome the biologist and medical-centered logic of hematologist-centered care.

From the second half of this year, in the context of budget cuts, social worker Maria Cândida Queiroz took over the coordination of the PNA-IPDF. Then, the political crisis escalated, given the political coup that culminated in the impeachment of President Dilma Rousseff in 2016. The economic crisis and political instability were the hallmarks of this period. During Maria Cândida Queiroz's administration, a movement was made to strengthen the Policy for the Care of People with SCD through technical advisory visits to all the states and regional workshops that aimed to assist in the implementation of the state's Care Lines, respecting local specificities, involving management, services, and the social movement to include SCD in existing health networks.

That same year, the social movement of people with SCD joined the CGSH in demanding that SCD be included on the list of pathologies eligible for bone marrow transplantation (BMT). In Brazil, some 1,000 people have been cured by BMT in university research centers. The Ministry of Health/MS responded favorably to the publication of the Ordinance. However, it restricted the age at which the procedure could be performed, which continued to mobilize the associations to adjust this document. BMT has also been touted as the only way to cure SCD.

The election of Jair Bolsonaro as President of the Republic is a significant turning point and represents a step backward in SCD-related

health policies. The rise of an extreme right-wing government in Brazil (2019-2022), represented by the Bolsonaro government, was a severe moment of setbacks or stagnation in public policies, characterized by the paralysis or continuous underfunding of projects and programs, alteration/revocation of several legal frameworks due to the implementation of a movement to dismantle programs, policies, or technical areas as a government program strategy, with a strong ideological slant, especially in actions focused on gender and racial equity, or in defense of vulnerable populations or specific groups. In the field of health, for example, the National Health Council (CNS) denounced the withdrawal of resources from the Unified Health System (SUS) for 2023 in a letter sent to the United Nations (UN) Health Rapporteur²⁴. Compared to the 2022 budget, which was equivalent to R\$ 172.60 billion, the budget for the Ministry of Health in 2023 was reduced by R\$ 22.7 billion. The CNS also denounced that these losses would reach R\$60 billion if the resources sequestered by the "Spending Cap"²⁴ were considered and was another challenge to the third term of President Luís Inácio Lula da Silva's government.

2) Historical and current neglect and the impact of racism on the history of SCD in Brazil

Racism in Brazil is structural and systemic, engendered in institutions and subjectivities. It is a "process in which conditions of subalternity and privilege are distributed among racial groups and reproduced in the political, economic, and daily relationships spheres"²⁵.

The case of sickle cell disease illustrates the relationship between racism and negligence. Prevalent in the Black population, it was first reported scientifically and widely in 1910 by Dr. J. B. Herricks, who, in the journal *Archives of Internal Medicine*, described the case of a 20-year-old Black student, born in Jamaica, who had a hitherto unknown clinical and hematological condition: episodes of jaundice, palpitations, and difficulty in performing physical exercises, and a blood test revealed anemia and several elongated "sickle-like" cells²⁶. This fact introduces SCD to the scientific and research world. It traces a roadmap of non-assistance that lasted until 2005 in Brazil when the National Policy for the Comprehensive Care of People with Sickle Cell Disease was launched through Ordinance n° GM 13/95/2005.

This gap of almost a century between the “discovery” and the first public policy expresses an important social invisibility component because some lethargy was observed in the field of care amidst incessant development of scientific research on the subject, which leads us to wonder how scientific knowledge can end up not being used as public health policy²⁷. This *sine qua non* condition was presented to us by Naoum, who, in the preface to his book *Sickle Cell Disease*²⁸ (Our free translation of the Portuguese title), makes us reflect: “It was precisely the Black African who, by suffering from a chronic and painful illness such as sickle cell disease, contributed with his pain, blood, and early death to the most important scientific knowledge about the biochemistry, physical chemistry, genetics, and molecular biology of proteins. Despite all the progress made so far, Black people worldwide, especially Black Brazilians, have not benefited from the scientific and technological achievements made with their blood...”

Cyclically, racism can be considered the beginning, cause, and outcome that explains why Black people have the worst health indicators. In this sense, SCD history intertwines with that of many other diseases that are not prioritized because they affect a disadvantaged section of the population. A disease is considered neglected when it is not prioritized by health policies despite its social relevance, either because of the number of people it affects, its lethality, or the level of suffering it causes. These are often diseases that evoke a “discourse of segregation, suburbs, and oblivion, whether by the pharmaceutical industry, governments, or health systems”²⁹, contributing to cyclical neglect.

Neglect can result from social invisibility because it is not recognized as a health issue. It can also result from apathy when the problem is recognized but needs to be given due importance. It can also stem from inaction or a lack of action to respond to the problem, which can also be due to incompetence in planning and conducting actions³⁰. Whether for one or more reasons combined, the production of negligence is closely linked to the low prioritization of issues and political disinterest.

We should recall that, although the causes are structural, neglect in health does not just happen; it is caused by invisibility, apathy, inaction, or incompetence³⁰. There is a process of producing neglect, encompassing both structural dimensions and the role of agency, omission as a social action, or non-recognition as a political project. It

is not just neglected diseases but neglected people, populations, territories, and communities through complex historical and social processes.

Prevalent in the non-white population, SCD is found worldwide, with a higher concentration in some African countries (sub-Saharan Africa) and India. Approximately 300,000 children are estimated to be born with the disease every year³¹. However, these children’s survival and quality of life will depend on their country’s economic and social development, the availability of healthcare, and the conditions to access this care. The disease has an increasing prevalence globally. However, in developed countries, the number of people with the disease is mainly due to increased life expectancy from health interventions³¹. Despite progress, infant mortality among children with SCD is still a reality in impoverished contexts³¹.

Almost all health organizations and governments³² have ignored SCD. The author affirms that the following characteristics justify this argument: (1) This disease affects millions of people worldwide; (2) It has a more significant impact on impoverished populations; (3) It has a high morbidity and mortality rate; (4) It can act in comorbidity with other equally lethal diseases; (5) It is simple to diagnose and (6) It can be treated with low-cost options. There is still insufficient data on mortality from SCD in Brazil³³⁻³⁵, and more studies addressing this population’s aging and the most frequent complications are required.

There is a need to draw the attention of federal funders, legislators, lawyers, researchers, and health decision-makers to work collaboratively and in partnership to reverse the impact of years of structural racism on SCD³, as seen in the disparate distribution of resources, as in the US scenario, where the allocation of resources for cystic fibrosis is much more significant for a smaller white population than the massive number of Black people with sickle cell disease. The comparison with cystic fibrosis is pertinent because it is a hereditary, progressive, and life-threatening disease associated with reduced quality of life and a shorter lifespan, just like sickle cell disease. However, cystic fibrosis mainly affects white Americans, in an equivalent proportion of one-third less than the Black population. However, it receives 7 to 11 times the research funding per patient, resulting in disparate drug development rates. In practice, this means the approval of four drugs for sickle cell disease and fifteen for cystic fibrosis by the Food and Drug Administration. The authors conclude that it is necessary to ana-

lyze the role of race and racism in this persistent (mis)allocation of resources.

3) The current situation of sickle cell disease: concern and hope

The pandemic has contributed to the recrudescence of multiple crises, mainly affecting people with SCD, increasing the risk of illness and death. As it is a systemic disease, which entails a greater risk of inflammatory processes, we expect an increased risk of people with SCD developing severe COVID-19 cases with a more significant likelihood of renal, neurological, and cardiovascular complications³⁶, and this can be seen in the National Immunization Plan, which lists sickle cell disease as a priority comorbidity for vaccination. Hence, there is a need to prioritize immunization in people with SCD since it is the only effective strategy to avoid contamination or even mitigate the COVID-19 effects. By unraveling the imbrications of SCD in the previous sections, it is easy to understand that any intervention decontextualized from the ethnic-racial issue gives rise to unfair handicaps, which criticize the criterion for prioritizing vaccines by age group and identified comorbidities. Priority access to vaccines for people with SCD is a legitimate demand from the perspective of equity and, above all, the right to preserve life.

The lesson learned is that we need to guarantee a comprehensive, multiprofessional approach to people's health, comprising community health workers and health teams consisting of doctors, nurses, dentists, biomedical doctors, pharmacists, physiotherapists, occupational therapists, psychologists, social workers, educators, and all the other professions that qualify for the well-being of people with SCD, from health education,

health promotion, prevention, and treatment to rehabilitation and health surveillance.

Brazil lacks a monitoring panel for SCD cases. For example, the US experience with SCD and COVID-19 cases led to a worldwide collaborative network of health professionals who treated people with SCD and recorded and monitored confirmed COVID-19 cases, which allowed them to monitor cases and, above all, compile data and information for developing actions and decision-making.

Besides the challenges posed by the pandemic, many other impasses hinder comprehensive care for people with SCD inside and outside the health sector. The country's current severe economic crisis directly affects SUS, and underfunding is a reality. Joice Aragão returns to the Ministry of Health in 2023, now as general coordinator of blood and blood products, and with her, a whole group of health professionals, social control, managers, and researchers, with the hope for progress in implementing the PNAIPDF.

Final considerations

Sankofa is the guiding principle of our analysis. *Âgô*, we ask permission to access the difficulties, mistakes, and success of a trajectory in which we, the authors, are also part of a network of mobilization and struggle, whether as researchers, health professionals, managers, or family members of people with Sickle Cell Disease. In conclusion, when reflecting on the paths to be followed, we point out that the work fronts should be taken up by managers, health professionals, universities, and researchers who value permanent dialog with people with SCD and their families.

Collaborations

All authors collaborated equally in all stages of preparing the manuscript.

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