

Article

Sickle Cell Disease in Bahia, Brazil: The Social Production of Health Policies and Institutional Neglect

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Abstract: A disease is considered neglected when it is not given due priority in health policies despite the social relevance of that disease, either in terms of the number of individuals affected by it or its morbidity or mortality. Although the causes are structural, neglect in health does not occur in a vacuum. In this paper, we explore how sickle cell disease (SCD) is constructed and neglected in Brazil, based on insights from our long-term participatory qualitative research in the state of Bahia. We present five overarching themes relevant to the social production of SCD, and associated health policies in Brazil: (1) The achievements and setbacks to overcome neglect in SCD, (2) Continuity of comprehensive SCD care; (3) Social movements of people with SCD; (4) Biocultural citizenship; and (5) Academic advocacy. We conclude that it is insufficient to merely recognize the health inequities that differentiate white and black populations in Brazil; racism must be understood as both a producer and a reproducer of this process of neglect. We conclude with a set of recommendations for the main SCD stakeholder groups committed to improving the lives of people living with SCD.



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‘O contrário de negligência é cuidado. Cuidado, questão central para saúde. Então, poderíamos perguntar: como encontrar cuidado em uma sociedade baseada na notoriedade e no (re)conhecimento? Se eu não sou reconhecido, não existo para as políticas públicas, não me comunico nem se comunicam comigo, não apareço em lugar nenhum, não recebo cuidado. Eu sou negligenciado, eu adoço.’ [1] (p. 7)

‘The opposite of neglect is care. Care, a central issue for health. So, we could ask: how to find care in a society based on prominency and (re)cognition? If I am not recognized, I do not exist for public policies, I do not communicate with others or vice versa, I do not appear anywhere, I do not receive care. I am neglected, I get sick.’ [1] (p. 7)

1. Introduction

In this paper, we explore how sickle cell disease (SCD) is constructed and neglected in Brazil based on long-term research in the state of Bahia. We will first briefly discuss the concept of ‘neglect’ in health, followed by a reflection on the group of diseases commonly referred to as ‘neglected tropical diseases’, before we frame how we traced the social production of SCD and its associated health policies in Brazil.

1.1. The Production of Neglect in Health

A disease is considered neglected when it is not given due priority in health policies despite the social relevance of that disease, either in terms of the number of individuals affected by it, or its degree of morbidity or mortality rates. Although the causes are structural, neglect in health does not occur in a vacuum. The production of neglect entails

a process that encompasses the structural dimensions, the role of the agency, omission as a social action, and a failure to recognize it as a political project [2].

The United Nations' agency responsible for international public health, the World Health Organization (WHO), brings together a diverse group of 20 diseases which it defines as 'neglected tropical diseases' (NTDs), as follows: 'NTDs are mainly prevalent in tropical areas, where they mostly affect more than 1 billion people who live mostly impoverished communities. They are caused by a variety of pathogens including viruses, bacteria, parasites, fungi and toxins. These diseases cause devastating health, social and economic consequences to more than one billion people' [3].

The label 'neglected tropical diseases' may well reflect a colonialist conception and a geographic determinism that could already have been overcome, by thorough reflection on and adjustment of the label [4]. The term '*tropical*' marks out a certain geographical area: 'the tropics' as a site of diseases associated with the climate, vegetation, and even the culture. This represents the vestiges of a certain colonial mentality and a political interest in dominating and perpetuating subordination. The existence of these diseases is the result of political, economic, and scientific neglect, related to the lack of engagement with marginalised communities [5]. Indeed, the persistent underfunding of research, the political disinterest, and the lack of effective public health policies are some of the factors contributing to the production of neglect [2]. It is crucial to understand the existence and perpetuity of neglected diseases and their relationship to poverty [3], exploitation, and the subordination in colonial historical process and dominance of capitalist societies. Although the most visible facet of inequality is injustice in the access to and distribution of material goods and basic health services, it is important to recognize that this is intrinsic in power relationships and dominance in capitalism [6].

1.2. Framing the Problem

Why do some diseases receive more attention from health policy makers and researchers, are allocated vast resources in health systems, and are granted awards from funding agencies? Which factors are decisive in such prioritization of diseases, often to the detriment of other diseases, many of which are more prevalent in certain regions? What are the legacies of colonialism in public health policies? These and other questions should be central in the debate on the production of neglect in health and are crucial to consider in relation to health inequities and the impact of such inequalities on individuals with neglected diseases.

In 2006, SCD was recognized by the WHO as a global public health problem [7] but is currently not included in the WHO's list of NTDs. It is pertinent to question why SCD has not been recognized as a neglected disease. In this paper, we examine the production of neglect through a reflexive analysis of SCD in Brazil, where structural racism is a major determining factor in the production of SCD-related neglect, recognizing the intersections between institutional racism and the production of neglect. Our main contention is that there is urgent need for a focus on the political, scientific, and economic determinants in the production of neglect of certain diseases and how (the lack of) prioritisation for these diseases reinforces and reproduces such neglect. As we will explain below, SCD is prevalent worldwide, particularly in the poorer regions of the world and especially in black populations. Structural racism has been assumed to be one of the determinants of the production of neglect, through the intersections of certain social markers, notably race, social class, and gender.

1.3. Sickle Cell Disease

'Sickle cell disease' (SCD) is an umbrella term for clinically severe sickling syndromes, characterized by the presence of the gene for hemoglobin S (HbS). SCD is thus not one single disease, but refers to a collection of genetic blood disorders characterized by structurally abnormal hemoglobin variants. The name derives from the elongated sickle (crescent) shaped erythrocytes which disrupt the blood flow in small vessels. The first description of

the disease occurred in 1910 when sickled erythrocytes were identified [8]. In 2022, SCD can manifest in some people as an acute and life-threatening condition, and for others, SCD can be managed as a chronic condition which requires comprehensive healthcare throughout the life course [9]. This may include specialized care for acute events or complications cur [10], especially derived from two processes: severe anemia and vaso-occlusion [11].

The diagnosis of SCD is simple as there are several techniques that can identify abnormal hemoglobin S (HbS). Such identification happens through a blood test called electrophoresis. An early diagnosis is recommended since the first symptoms appear around six or eight months of life [12]. Such early diagnosis allows for initiating health care, including special immunization and the introduction of oral penicillin. The impact of early diagnosis is clear, as mortality was reduced from 25% to 3% and it also reduced the severity and frequency of complications [13,14].

The main clinical features of SCD are acute pain and manifestations, including infections, anemia, and organ damage. Sick cell anemia is the most common and severe form of SCD and is characterized by the double presence of the gene S [8], affecting over 30,000 individuals in Brazil [10]. It can lead to many complications, including chronic anemia, vulnerability to severe infection, acute painful crises, stroke, kidney problems, chronic pain, cardiopulmonary complications, leg ulcers, and osteoarticular lesions [15].

SCD is one of the most common genetic disorders worldwide [16]. Projections suggest a 30% increase in the number of individuals with SCD by 2050 [17]. This growth is due to the implementation of healthcare actions focused on the disease that have significantly increased life expectancy [17,18]. The highest prevalence is in certain countries in Africa and in India [7]. As many as 300,000 children are estimated to be born with SCD annually [19].

The WHO estimates that SCD is the base cause in 15% of cases of child mortality in African countries [15]. Early childhood mortality is as high as 50 to 90% in low-and middle-income countries, and this has not changed since the 1970s [20]. The survival and quality of life of children born with SCD depends on the socio-economic context of the country in which they live, on the presence or absence of (access to) healthcare [7].

Although SCD is recognized as a severe global health problem, experts argue that little effort has been made to manage the condition from a global perspective [19,21,22]. This is so despite the fact that SCD treatment is available and inexpensive [22]. There have been calls to add SCD to the group of NTDs. Ware et al. (2013), for instance, argue that, although it is not transmissible, sickle cell anaemia (SCA) should be considered neglected [23]. He puts forward a compelling argument for SCA, which has a worldwide prevalence, to be considered a neglected disease because (1) it has been ignored by almost all health organizations and governments, (2) the most disadvantaged and impoverished communities carry the highest burden of SCA-related morbidity, comorbidities with other life-threatening conditions, and (3) a simple diagnostic test and inexpensive treatments are available for SCA.

1.4. The Context of Healthcare and SCD in Brazil

Of the countries that make up the Latin America and the Caribbean region, Brazil has the greatest number of cases of neglected 'tropical' diseases, and these diseases constitute a public health problem in the country [24,25]. The Brazilian health system consists of a hybrid public-private structure formed by a complex network of service providers and purchasers distributed in three subsectors [26]:

1. The public subsector (SUS)—services are financed and provided by the state in multiple levels (federal, state, and municipal);
2. The private (for-profit and non-profit) subsector, financed in various ways with public or private funds;
3. The private health insurance subsector, containing different forms of health plans, insurance premiums, and tax subsidies.

Based on the principles of universality, integrity, and social participation, the SUS was the most important output of the Brazilian Health Reform Movement in the 1980s. This

has its basis in the 1988 Brazilian constitution, which recognizes health as a citizen's right and a duty of the state. After its implementation, Brazil became the only country in Latin America with a universal health system [27]. We agree with Paim et al. [26,28] that, 25 years since the creation of the SUS, many advances have been made in the Brazilian health system, including improved access to several health services, institutional innovations (such as a substantial decentralisation process which gives the municipalities responsibility and resources for more effective local health services), and social participation in health policy making and accountability.

Despite these improvements, there are still many challenges in the Brazilian health system, especially the fragmentation of policies, underfinancing, the complex relationships between the public sphere and the market, and the weaknesses in regulatory processes and persistent inequalities in health [29]. While many countries with a universal health system spend between 7% and 8% of their GDP on public health actions and services, Brazil's expenditure was under 4% of the country's GDP in 2012 [30]. The inequity with regard to the access, availability, and quality of health services reflects Brazil's deep regional and social inequalities. The greatest challenge in the Brazilian health system is indeed to improve universal and equitable coverage of quality health services. The situation became worse in recent years. Unfortunately, many achievements and advances suffered significant setbacks after the impeachment of President Dilma Roussef and the imposition of austerity policies. As Paim [26] (p. 1794) put it: 'the greatest challenge facing the SUS is political: the SUS must now be guaranteed its political, economic, and scientific and technological sustainability'.

Among Latin American countries, Brazil leads in the number of new-borns annually affected by either SCD [31] (called *Doença Falciforme* in Brazil) or HbAS (sickle cell trait). More than 6000 newborns are born annually in Latin America, and approximately half are born in Brazil, with the estimation of approximately 2500 children born with SCD each year in Brazil [11,32]. The sickle cell trait (HbAS) is largely present in the population of Brazil, with the estimation of 2 million Brazilian carriers of the hemoglobin S [10].

In Brazil, and in other countries in North and South America, SCD is more prevalent among black populations, and within Brazil, SCD is even more prevalent in regions with higher concentrations of black populations [31]. This process can be explained by the three-hundred-year slave trade in Brazil and the conformation of a demographic profile of Brazilian society with a high presence of African descendants [32].

The state of Bahia concentrates higher incidence rates of SCD [13]. Concerning the sickle cell trait, an incidence of 1 per 17 live births is estimated in Bahia, 1 per 20 live births in Rio de Janeiro, and 1 per 30 live births in Minas Gerais [33]. Bahia also has the highest incidence of the disease [13], 1 per 650 live births [33]. In a region of Bahia, Recôncavo Bahiano, there are municipalities with an even higher incidence, 1 per 324 live births, in a region with a population of predominantly African ancestry [13]. Despite these high prevalence rates of SCD in Brazil, there are still very few studies focusing on the characterization of the socio-demographic profile of SCD patients in the country.

1.5. A Brief Historical Overview of the Production of SCD-Related Policies in Brazil

In 1947, Dr. Jessé Accioly, a professor at the School of Medicine of the Federal University of Bahia, identified SCD in Brazil and its genetic inheritance mechanism. The first public policy, the Sickle Anemia Program (*Programa de Anemia Falciforme—PAF*), aimed specifically at SCD, was only put forward five decades later, in 1996. The lack of resources meant it remained a proposal and went no further [34]. As with most health policies, the implementation of a SCD-related policy was far from straightforward. Such inaction and silence regarding existing health issues are forms of neglect.

An important landmark occurred in 2001 at the WHO's World Conference against Racism in Durban, where South African and Brazilian antiracism activists were present, with a prominent role played by black women [34]. As a result, affirmative action policies gained greater visibility and the Brazilian government recognized the need for anti-

discrimination policies [34]. SCD was an important focus for the anti-racism social movement. When the Black Movement included SCD in their agenda, it was clear that the first step was to fight for its recognition. An interview we conducted in 2015 with the physician who managed the national SCD program revealed details of these challenging times, as she recalls:

There were a lot of complaints, a lot of suffering, a lot of anxiety, a lot of complaints about the lack of knowledge about the disease, about the difficulties people faced. People with SCD did not feel supported. They felt abandoned, as if the Ministry and the health system left them unassisted. They felt left behind, as if the state, the public office, was not represented in the Unified Health System, lacked to engage with the population, the SCD population (Manager of the national SCD program between 2005–2015)

In 2001, SCD was included in the National Program for Newborn Screening for hemoglobinopathies (Ministerial Ordinance 822 of June 6, 2001) [12]. Nevertheless, the resolutions established in the document were only implemented in 12 of the 26 Brazilian states [35]. It was only almost a decade later, in 2010, that all the states had implemented newborn screening for SCD. This delay, in many states, had an extremely negative effect on the early detection of the disease and on the quality of life of people with SCD. Another manager of the national SCD programme reflected on this period during one of our interviews in 2015:

In terms of the SCD, there is still a great deficit, a great lack of visibility in information systems. We always worked on action planning, based on the data delivered by newborn screening. Newborn screening has been important, highly significant for [the mapping of] SCD, as it revealed this bigger picture. But only data from the new births are collected, only from children. Data from adults, from those who were diagnosed late are still lacking: who are they, how many are they, how are they? (Manager of the national SCD programme between 2015–2018)

In the same year, 2001, the National Federation of People with SCD (FENAFAL) emerged as a national health activist group. In the state of Bahia, the State Association of People with SCD (ABADFAL) was also created in 2001 by the parents of a young girl with SCD.

The social movement of people with SCD played an active role in the creation, formulation, and monitoring of policies towards people with SCD at the national, state, and local level. The influence of this social movement has increased over time. In 2005, there were eight associations of people with SCD in Brazil; to date, there are 45 associations. This is not only about the representative aspect of these associations, but also about their leading role in the development of policies and the continuous action to implement them. Even in the symbolic plan of the social representations of the disease, the movement of people with SCD carries out powerful transformations, as one interviewee explained:

The associations began to consider themselves as individuals. Before, they were persons with sickle cell (falcemics) associations—you will still find associations with that name. ABADFAL constructs an idea. Ideologically, ABADFAL makes a political intervention in the speech of the user, of the person with SCD in Brazil. They primarily think of escaping the label, the stigma. Because when we spoke of a SCD patient, this person was then called a ‘falcemic’, and the listener would immediately associate with someone who was not going to live long and would die early. When we intend to escape stigma, we have ideas. When I talk about people, I am talking about individuals, someone who is building their own history, which cannot be determined by the disease. (Former coordinator of the State Association of People with SCD)

Until 2004, ANVISA, the National Health Surveillance Agency, was in charge of the treatment of hemoglobinopathies. This was problematic, as ANVISA is not responsible for health care. That same year, the Ministry of Health launched the SCD Program and

designated a specific coordination for SCD and other hemoglobinopathies (General Coordination of Blood and Blood Products—CGSH). The creation of this general coordination was extremely important in order to gain political traction in the Ministry of Health and support the development of a care policy. It also expanded the participation of organizations of people with SCD and their influence on health policies. The National Federation of People with SCD (FENAFAL) gained a seat in all meetings for the organization of a national policy on SCD, establishing an intensive dialogue with the Ministry's managers. Also in 2004, a first national seminar on the health of black population groups, in which SCD appears as a key demand, was convened. The resulting publication focuses on issues such as racism, health-specific aspects, and policies targeted at ethnicity-related diseases. SCD is referred throughout the document.

It was in August 2005 when the Ministry of Health launched the 'National Policy of Comprehensive Care for People with Sickle Cell Disease and other Hemoglobinopathies' with the basic guidelines for health care [34]. One study participant reaffirmed the importance of this document in a 2015 interview:

Until 2005, there was no SCD in the Ministry of Health. There was no document ... like a directive that tells you what it is all about. Do you understand the problem? I mean, we overcame that... Do you understand the difference? The neglect is not recent. I think today, thanks to the social movement, we broke a barrier. (Manager of the national SCD programme between 2005–2015)

In 2015, Maria Cândida Queiroz, former coordinator of the Municipal Program for Health Care for People with SCD (PAPDF) of Salvador, Bahia, assumed the coordination of the National Policy of Comprehensive Care for People with Sickle Cell Disease and other Hemoglobinopathies. This was a great accomplishment for the social movement since she was also the mother of a girl with SCD and one of the founders of ABADFAL, with a great history of activism. The publication of the document 'Sickle Cell Disease: Basic guidelines of care line' (*Doença Falciforme: Diretrizes Básicas da Linha de Cuidado*) in 2015 is considered a great accomplishment in national health policy [36]. The document was published as an official directive to establish the flow of care and strategies for overcoming the logic of assistance focused on the haematologist, and for the inclusion of SCD in the comprehensive care network.

That same year, the social movement of people with SCD joined the Coordination of Blood and Hemoglobinopathies in the demand for SCD to be included in the list of diseases eligible for bone marrow transplantation (BMT). The growing prevalence of the procedure in Brazil represents hope to individuals with SCD, since it is the only proven cure for the disease [37–39]

In 2018, the election of Bolsonaro as president represented a setback in SCD-related health policies. The Blood and Hemoglobinopathies Coordination no longer had a person responsible for the National Policy of Comprehensive Care for People with Sickle Cell Disease, nor was a budget ringfenced as had been the case in the past decade. In 2019, a document about a National Policy for Rare Diseases was launched by the Ministry of Health. This mentioned SCD as one of the 'rare' diseases in Brazil. There was a strong reaction from SCD activists, researchers, and health professionals, claiming that such a statement was a great setback in the awareness of SCD as a public health problem in Brazil. A couple of months later, the document was removed from the website of the Ministry of Health and a modified version was uploaded, with no mention of SCD.

2. Methods

2.1. Longitudinal Qualitative Research and Activism on SCD in Bahia, Brazil

The insights on which this paper is based are drawn from two participatory research projects on SCD, each with nested qualitative research projects. The SCD research teams were comprised of researchers, undergraduate and postgraduate students, SCD community actors, and health policy stakeholders. This SCD research began in 2009 at the Federal University of Bahia, Brazil, with fieldwork in the state of Bahia. The overarching aims

of this longitudinal research are threefold. Firstly, to examine the challenges faced by people with SCD and their families, to map out their needs and the therapeutic itineraries as well as the contexts of vulnerability on an individual, community, and institutional level. Secondly, to map out the barriers and facilitators to receiving SCD healthcare, and the potential of the healthcare system in the city of Salvador, Bahia, Brazil. Thirdly, to contribute to improvement in quality of life and comprehensive health care for people with SCD.

These aims were met through different projects. In this paper, we use insights from two projects in particular. The first project, *Accessibility and equity in the primary health network from the perspective of the black population*, was led by Leny Trad between 2009–2015. We focused on the different perspectives on the needs of people with CSD and their families, as well as on the political dimension of the municipal health care programs and their role in the diagnosis and management of CSD. The second project, *Vulnerability, Therapeutic Itineraries and Comprehensive Care for Chronicity: a focus on Sickle Cell Disease (SCD) and Chronic Myeloid Leukemia (CML)*, was led by Clarice Mota between 2016–20. This addressed these main issues: therapeutic itineraries, daily life and experience with SCD and CML, and mapping out social and institutional support networks.

The first project, *Accessibility and equity in the primary health network from the perspective of the black population*, was carried out in the Sanitary District of Liberdade, involving a primary health care unit, which was the local reference of health care for SCD. The study participants were recruited in the waiting room of this unit: we invited them to participate and provided contact numbers if they wished to participate in an interview. We conducted seven interviews, including individuals with SCD and family members, who were interviewed in their homes. All identified themselves as black or brown (*pardo*). We also conducted five interviews with healthcare professionals (doctors, nurses, social workers, pharmacists, and community health workers) and four focus groups: with healthcare professionals, with community health workers, with young people, and with older adults.

From the start, we involved all stakeholders in the SCD community to mediate the dialogue among all health actors in order to strengthen the health care network for people with SCD. One very effective strategy was World Coffee, which consisted of workshops to articulate and activate the network. In the first workshop, we engaged in activities which promoted the interaction of the different health institutions responsible for health care, and we carried out a brief evaluation of the level of collaboration between these institutions. After the first World Coffee, we asked that each institution complete a form including information on the type of care they provided for people with SCD, in terms of capacity and services offered. During the second workshop, we had two main activities: (1) discussing the challenges of developing a network of care for SCD (what role would each institution play in the network); and (2) a discussion around three cases of individuals with SCD and their health itineraries. We used interview data and discussed responsibilities for the problems we identified in those three cases.

Along with the meetings and activities with stakeholders, we invited them to participate in the research. Over the first few years, eight stakeholders were interviewed: the former director of the Association of Parents and Friends of the Exceptional (APAE); two former coordinators of ABADFAL (Bahian State Association of People with SCD); two former National Coordinators of the National Policy of Comprehensive Care for People with Sickle Cell Disease (PNAIPDF); the former municipal coordinator of the Municipal Health Care Program of Salvador (PAPDF); the current coordinator of the National Federation of People with SCD (FENAFAL); and the former director of the Primary Health Care Unit of the Sanitary District of Liberdade.

The second project, *Vulnerability, Therapeutic Itineraries and Comprehensive Care for Chronicity: A Focus on Sickle Cell Disease (SCD) and Chronic Myeloid Leukemia (CML)*, involved adults who lived with SCD who were interviewed in the University Hospital where there is an Ambulatory for Hemoglobinopathies. We conducted a total of twenty-seven interviews. In terms of the social profile of the participants of this research, fifteen were women and

eleven were men, whose ages ranged from 18 to 67 years. Fifteen identified themselves as brown (*pardo*), eleven as black, and one as white.

It is important we emphasize here the contributions of the research assistants, and graduate and undergraduate students throughout both projects. The group of research assistants included four people who live with SCD and also the father and mother of a girl with SCD. They were all partners in the project development and co-authors of the publications. The research generated twelve dissertations on a range of SCD-related topics, including gender and SCD, reproductive rights of women with SCD, comprehensive care for adults with SCD, barriers to SCD-treatment in hospitals, SCD-mortality in Salvador and the role of social movements in health care.

2.2. Collaboration with SCD Stakeholders

In this section, we outline our approach to our close working relationships with both community actors and health policy makers during our SCD research. We always invited the Bahian Association of People with SCD (ABADFAL) to participate in discussions at all meetings and workshops. In turn, we, the researchers, were invited by them on many occasions to provide feedback from our research and discuss our findings. The more we listened to the trajectories of suffering and the struggles people with SCD in Bahia faced, the more we reached a profound understanding of the complexity of SCD in Brazil.

Another strong partnership was the one with the Municipal Health Program of Salvador (PAPDF). This partnership reflected our objective of having a direct and concrete impact on improving care for individuals with SCD and their families. We participated in several training activities for healthcare professionals, which we co-developed with the municipal program. As a result of World Coffee, the PAPDF started having monthly meetings with the services involved in SCD healthcare delivery for SCD and invited us to participate. Through this collaboration, we were able to identify and robustly map out the challenges around (a lack of) communication and smooth referrals between different levels of care, which were failing to meet the healthcare needs of people with SCD.

A final important engagement was our participation, over the years, in two annual events promoted both by the Municipal Health Program and ABADFAL: the Week of Awareness of SCD, every October, and the International Day of Awareness of SCD on 19 June. The events involve health education strategies, blood testing, workshops, and leisure activities held in different localities. Our research teams would support the events, give academic visibility to them, and engage students in these activities.

In 2015, the National Sickle Cell Disease Program of the Ministry of Health asked us to organize a national training course about SCD for over 1500 primary care professionals from twenty-six Brazilian states. The 'Online Course Comprehensive Care & Sickle Cell Disease', Institute of Collective Health at the Federal University of Bahia, by Clarice Mota and Liliana Santos. This course was a partnership between academia, health policy makers, and the associations of people with SCD, with the main focus of the training of health care professionals being around the principles of the Universal Health Care System (SUS) as applied to comprehensive care for people with SCD. The course was designed to provide education on SCD, not only on the physiological, but also the psychosocial aspects of living with SCD.

The achievements of the twelve years of investment in research, advocacy, and technical cooperation in SCD can be seen as parts of one overarching participatory project, based on the collaboration between universities, activists, people with SCD, and their family members, as well as municipal, state, and national stakeholders. It resulted in sixteen publications, mostly in Portuguese, so that the content is accessible to health professionals, stakeholders, and communities in Brazil. During the COVID-19 pandemic, we contributed to a technical note around the implications of COVID-19 in individuals with SCD who are more vulnerable to cardiorespiratory complications [40].

2.3. Decolonial Approach to Community Participation

We strongly believe that community–academia partnerships should include all institutions responsible for SCD health care, as well as social movements advocating for people with this disease. That is why it is so important to use the methodology of community-based participatory research. Such partnerships can tackle the SCD health inequities and to enhance the understanding and visibility of SCD in Brazil.

This methodology also seeks to overcome hierarchical and fragmented perspectives, reaffirming the importance of connecting the university with government and non-governmental key stakeholders. In operational terms, it is a process where all partners are involved in understanding the problem identified, as well as in the process of planning, implementing, describing, and evaluating the investigation. It also includes promoting changes in social representations and stimulating the social participation within the community [41]. The results are accomplished through a collaborative approach between researchers and key stakeholders.

3. Results and Discussion

In this section, we bring together five overarching themes relevant to the social production of SCD, and the associated health policies, in Brazil:

1. Achievements and setbacks: the struggle to overcome neglect in SCD;
2. Continuity of comprehensive SCD care;
3. Social movements of people with SCD;
4. Biocultural citizenship;
5. Academic advocacy.

3.1. Achievements and Setbacks: The Struggle to Live with SCD

One of the landmarks in the implementation of an SCD health policy in Brazil was the inclusion of SCD in the neonatal screening program in 2001, which allowed early diagnosis and care [13]. As with all procedures in the Brazilian Unified Health System (SUS), newborn screening is free of charge. It has proven to be a very effective health policy [10,13]. Unlike in other countries, newborn screening in Brazil is a universal health policy and its coverage is usually high, ranging from 85 to 87% [13,42].

The importance of such early screening is not only confirmed by the medical literature, but it was a recurrent theme in our data, the quote below is one example:

The sooner you diagnose it [SCD], the better the treatment will be, you know? That sometimes we see some complications in childhood, like . . . I think in one year I was hospitalized three or four times. So, if I was diagnosed early, my mother would be aware of the disease, which my mother didn't know. My father didn't know either, two ignorant people, let's say (36-years-old man with SCD)

In the second project, we interviewed 27 adults with SCD, and only six of them had received an early diagnosis through newborn screening. Ten of them were diagnosed during childhood, ranging from 3 to 10 years of age. One individual was diagnosed at 17 years of age and the other interviewees were diagnosed when they were already adults. Spending so many years with undiagnosed symptoms was painful and stressful for the person and their family, as was the case for this woman who was diagnosed when she was 40:

In the countryside my mother used to take me to pray a lot, she used to say: 'What is this?' (. . .) A lot of headaches. My father would say: 'Let's take it to see what's wrong', it was something that in the rural areas no one would ever find out what it was. (...) Since I was a little girl I had these crises, crises of feeling bad (. . .) There were days that I would arrive at work feeling very low, looking like I was going to faint. Every time I had a blood test: 'Ah, you have a little anemia, you have a little anemia', but we couldn't find out what was the cause of the anemia and through this test at the hospital we discovered it. For me it was a

great victory. At least I know what I have and I treat myself (60-years-old woman with SCD)

Another example of a man who was diagnosed with SCD at the age of 25:

I went to a doctor, I went to another one and nobody could figure out what was wrong with me, so, even so, I had to keep my job to be able to do my work, to keep me going, right? Even then I was sick, without knowing the cause of the problem, because I went to the doctor, I went for tests, at that time I had for several private tests, which my boss paid for, because I could not afford those. (. . .) Then this doctor there referred me here to the hospital, and here I came to do the test properly and they discovered that I had sickle cell anemia, and then I started the treatment. (52-years-old man with SCD)

The diagnosis can also come after a disruptive health problem that cannot be understood. This was the case with this 42-year-old woman, who only discovered that she had SCD at age 37, when she was suddenly paralysed from the waist down:

I tried to get up and could not walk. I lost all movement in my legs from the waist down. That is when I found out that I had lost 50% of the head of my femur. When the doctor started to run several tests, he discovered that all because of SCD. I immediately began treatment. That is why I am here in the hospital (. . .) Constant treatment. I already had two surgeries, and I am fighting not to have a third. All due to SCD. (42-years-old woman with SCD)

Another important landmark was when the Ministry of Health included, in 2011, the hemoglobin electrophoresis (SCD diagnosis test) in prenatal care routine. Two of our study participants received an SCD diagnosis during pregnancy, despite having previous symptoms.

My mother had a lot of pain. When she was young, before she got married and had children, she suffered a lot. She would go to the doctor, take medication, come home and it was no use. She had to go back again, and nobody found out what it was. (24-years-old woman with SCD)

I discovered I had SCD when I was pregnant. I did the test. My daughter was born with the trait. (37-years-old woman with SCD)

Although important, newborn screening is not sufficient to guarantee improvement in quality of life. Researchers in the Brazilian states of Minas Gerais and Rio de Janeiro investigated the mortality rate among children with SCD, comparing the ones born before and after screening for SCD, and concluded that this policy was not enough to reduce the mortality rate of children with the condition [43]. Adequate, continuous, and comprehensive health care is crucial to prevent not only SCD mortality, but also the sequels resulting from disease complications [17]. Simple preventative strategies, like penicillin prophylaxis and specific vaccines, can have a great impact [31].

It is important to recognize that high coverage of newborn screening in Brazil allowed us to produce data on SCD incidence, revealing the scale of the condition and its presence among the Brazilian population. Nevertheless, the lack of current data on the prevalence of the disease and the lack of a nationwide electronic database are indicators of neglect [32]. The health-related database in Brazil, available on a public platform called DataSUS, is adequate to allow numerous analyses to be performed on other diseases, but it fails to include SCD. The lack of national data on the number of individuals affected by the disease, their spatial distribution, sociodemographic profile, morbidity and mortality profiles, etc., may reflect a relative political disinterest. This is both the effect and the cause of invisibility, which, in a vicious circle, reproduces the idea that SCD is unimportant because it is not very prevalent, while also causing it to remain invisible because no data are produced.

3.2. Continuity of Comprehensive SCD Care

In terms of health care improvements for people with SCD, some achievements must be pointed out, in particular the incorporation of hydroxyurea treatment as part of the medical protocol in the Brazilian Unified Health System in 2013 (Decree n. 27, 12 June 2013). The progressive use of hydroxyurea has represented a significant increase in the quality of life of people with SCD, which is responsible for reducing morbidity and mortality [7,42,44]. The illness trajectory of our study participants revealed the positive impact of hydroxyurea: fewer painful episodes and shorter hospital stays, which was also confirmed by other studies [42,44].

If I had discovered this hydroxyurea before, I do not think I would ever go through what I have been through. Because God knows. My family knows what I have been through. I suffered a lot. Because the crises I had in the past. If I had these today, I would rather die. Because I preferred death. (. . .) Standing in a bed moaning in pain, grinding my teeth, twisting in pain and no medication, morphine, all kinds of strong medication and the pain did not go away. Today I am very well! (35-years-old man with SCD)

The hydroxyurea, the codeine and the dipyrone it is already helping me. I can stand it [the pain], do you understand? Before I was taking the medications I gave my family a lot of grief (. . .) I screamed a lot, I remember when I was little, I screamed a lot. After the prosthesis surgery I did physiotherapy, and after the doctor discovered this medication [hydroxyurea] I am carrying on with my life, right? I have a better quality of life (53-years-old woman with SCD)

Some of our adult study participants were in the process of evaluating the possibility of using the medication:

The doctors said that in order to avoid crises, so severe and so frequent, hydroxyurea is used to try to alleviate these crises. But with hydroxyurea, they say, there are side effects. That is what they say. (. . .) They said that, after tests, we will know if my body can support it. If my organs are all right, to know if I can really take it. (24-years-old woman with SCD)

The doctor said that she is ready to do some tests that will indicate if I can take hydroxyurea, because I have many crises (...) She said that hydroxyurea helps to fight any type of infection (...) But I have to do all the test first I have to know, she has to know how my kidney is, how my liver is, how everything is, in order to know if the medication is good for me or if it will not harm me (48-years-old woman with SCD, working as a teacher at the time of the interview).

As the interview excerpts above reveal, the use of hydroxyurea requires routine follow up blood and other exams. Some of these exams are not easily available in the public system (SUS), as mentioned by some of the study participants:

Examination in the SUS takes time, I do everything private because is faster. The ones I have here, I paid for all of them, because is faster. It was really difficult to get them at SUS. Sometimes the remedies are missing too, so I have to buy them all. (60 years old woman with SCD, from a small municipality in the state of Bahia, working as a house cleaner at the time of interview)

Despite the salary that my sister has . . . the benefit, practically we spend it all on investigations. Because most of us cannot get through the SUS and when it gets too long and end up missing the consultation deadline. And then you can never do it in SUS. And we end up spending a lot on tests. (24 years old woman with SCD, whose mother and sister also live with SCD, from a small municipality of Bahia, not working at the time of the interview)

In addition to a tiring routine for people living with chronic disease, geographical and economic difficulties limit the possibilities of ensuring continuity of care.

We're trying to start our life over, in a house with only two bedrooms, two small bedrooms, a small living room, a small, horrible kitchen. I am not ashamed to talk, my savings I spent everything on this disease. Sorry I talk, but it is a miserable disease, a disease that is destroying me. (40 years old woman with SCD, who lost 3 siblings with SCD, from a small municipality of Bahia, working as a teacher at the time of the interview)

The geographical difficulties were highlighted by the study participants who lived in rural Bahia. They mentioned challenges in accessing specialized care in their municipalities, especially haematology consultations and required clinical examinations.

We have to leave the house on Sunday, sleep in the secretariat to wake up and leave there at two o'clock in the morning. Then you come here [in the specialist unit] at six o'clock in the morning and then we need to wait all morning and the appointment is scheduled for one, but only three patients are seen [by the doctor]. Do you understand? (...) We sleep in the health department there. (...) For health transportation. That's it. Because the car leaves at two o'clock in the morning. (24-years-old woman with SCD, whose mother and sister also lives with SCD, not working at the time of the interview)

I have to come in the city hall car. When it [the car] is not available, I have to pay for it [transportation], right? It gets a little more complicated, but I have to come. The city is too slow in matters of transportation . . . to come to the doctor. Then the person has to plan first to organize the money to pay for a car. (32-years-old woman with SCD, working as a craftswoman at the time of the interview)

Dealing with SCD as a chronic condition demands a lifetime of adjustments. This also involves the family members. Sometimes the caregivers have to give up their jobs to care for their children. For instance, we interviewed a 37-year-old woman who has two children with SCD and could not work because she did not have someone to share childcare responsibilities with:

For me it is difficult, it becomes difficult because I do not go out and leave them... only if they are here with my mother. (...) When they have a crisis, when they have a fever, they have to have the right medication, understand? My family takes care of them, but nobody likes to go out with them, because they are afraid. I wish I could work, but I have no option. Because every time I get something to do, he gets high fevers. If they are hospitalized, I am hospitalized with them too.

Sometimes she has to leave the house to stay a long time in the hospital. There comes a time when she is nervous, she already comes with her problems (...) She has to leave home, leave her husband, leave everything. So sometimes they were like this, very emotional. It is important for the healthcare professional to see the other side of this family. Because it is not easy in a family with a chronically ill patient. It is not easy. (Pediatrician who worked at the reference service for people with SCD at the time of the interview)

This last statement highlights the role of mothers as caregivers and the burden they carry. The pediatrician added:

Usually the ones who carry the highest care burden, more than 90%, are the mothers. They know everything. They already know what should do for their child, to try to reduce the child's hospitalization. So they are more attentive.

Recalling the past, when she had a boy with SCD, a 60-year-old woman talked about her effort to understand SCD:

I did not know anything about SCD. I went to the library, because in the olden days there was no computer, you know. I went to the Central Library and asked the girls: 'Come here, is there a book about SCD?'

Her son, who is now 36 years old, also remembers the past and the difficulties he faced in continuing his studies despite the constant hospitalizations.

In my childhood it was painful. Like this... The school year started, in June it stopped, I was three years behind in my schooling. I was supposed to graduate when I was 18 and I graduated when I was 20. Because back then I had to stop. Monday, Friday, Thursday... every time I was in hospital. I remember that my mother stayed with me 24 h a day. Once I was hospitalized for 90 days, because of SCD, I think I was 7 years old. Thank God, today this is no longer the case.

In addition to these challenges, adults with SCD also face difficulties in maintaining their jobs during the periodic SCD crises.

We do not always wake up well, right? Sometimes with pain, sometimes feeling unwell, feeling sick, you know, that feeling of weakness, of fainting. But since you are providing a service, you cannot always afford to stay home and not go to work, right? I have worked as a domestic worker, but I cannot lead an ordinary life because of the disease. (33-years-old woman with SCD)

Sometimes the employer does not want people with health problems, you know. We miss work, I used to miss work because I had a crisis and I had to miss it, I could not work. So I went back to the rural area. Then I started to work in agriculture there, but then the illness increased, it increased more and more, then I got a disability pension (56 years-old-man with SCD).

A chronic condition such as SCD requires more research on how complications evolve over the life course [20], not only in clinical terms but also in the SCD-related psychosocial aspects.

The mothers are well oriented, so when they appear we feel this, but when the children get older, they end up having some kind of complication and then when they arrive, they arrive in a situation of hospitalization, chest crisis, those strong abdominal pains, in need of transfusion, you understand? So I have dealt with such complications, and we think: could we be avoiding this kind of thing? So I think that a basic level, well done, but we should also be reducing these complications in the future (Pharmacist who worked at the reference service for people with SCD at the time of the interview).

From the 27 interviewees of the second project, we observed sequels and comorbidities that can be attributed to the absence of proper health care across the life course. Of the comorbidities mentioned by our study participants, the most common were: hypertension (three individuals), retinopathy (four individuals), kidney problems (one individual), heart disease (two individuals), osteonecrosis (three individuals) and stroke (two individuals), diabetes (one individual) and leg ulcers (one individual).

I have many health problems, not only SCD. I have a heart condition. I have unstable angina pectoris. I have an eye problem, retinopathy. And now on top of that I have hearing problems—we are talking, and I am listening to you because you are here close to me. If you were over there, I wouldn't hear a word you said. I have high blood pressure. I take medicine for my blood pressure, medicine for my heart. And I had thyroid cancer. (48-years-old woman with SCD, working as a teacher at the time of the interview)

As per the statement above, the combination of SCD and other morbidities represents an extra burden for the person who lives with the condition and for their families, besides representing a challenge to the health system. This points to the need for a greater number of specialized care professionals, a greater increase in the number of hospital beds, and a greater capacity of the network to be prepared to serve and care for older people. It is urgent to rethink SCD and its health care model, in order 'to shift disease management processes from an acute care model to a chronic care model' [45] p. 604.

Comprehensive health care has long been a battle flag of SCD advocacy. Comprehensive SCD care should also include the primary care system, offering nutritional advice, therapeutic treatment, pharmaceutical advice, physiotherapeutic consultations, pediatricians, and general practice [12]. Even though this model of health provision is already established in the document *'Sickle Cell Disease: Basic guidelines of care line'* (2015), approved by the Ministry of Health, there is still a long road ahead until all Brazilian states provide comprehensive health care.

3.3. Social Movements of People with SCD

Both coordinators who managed the National SCD program since 2005 expressed the view that the social movement of people with SCD had an active role in the creation, formulation, and monitoring of policies towards people with SCD at the national, state, and local level. Here is how one of them explained this in a 2015 interview:

You may be shocked: 10 years ago there was no medicine with an International Code for Diseases (ICD) for SCD, folks! The disease ICD number and medicine are elemental for what? To be financed by the system, for the patient to get it for free. Hydroxyurea, the most important medicine in the treatment of SCD, had no ICD. The doctor had to use the ICD from another condition. This was not happening in the past century, but in this one. Do you get it? We have to deal with all these kind of things, actually within the Ministry, but at least we are now being seen. What I find comforting is that we became visible by our own efforts. This has to be credited to the Associations. We were all deep down, and these people have pulled us to the surface. (Manager of the national SCD program between 2005-15)

As mentioned earlier, in 2005, there were eight associations of people with SCD in Brazil. In 2022, there are 45 existing associations. In the last ten years, the partnership between the FENAFAL and the Ministry of Health has led to many outputs: seventeen documents were published in the form of manuals, brochures, and other informational materials for both the general population and health professionals; and fifteen directives were published, establishing clinical protocols, ensuring drug treatment, and access to diagnostic technologies.

In Bahia specifically, we highlight the prominent role played by the Bahian Association of People with Sickle Cell Diseases (ABADFAL) in the struggle for the right to comprehensive care, in the dissemination of knowledge and information about the problem among social support networks, in effective actions of social control, as well as in the empowerment of people living with the disease, whether in the condition of patient or caregiver.

SCD . . . that was really my world and that was it. I was the only one who was in pain, the only one running to the emergency room. After ABADFAL, I realized that there are others in situations worse than mine . . . I thought there was no life outside my world. ABADFAL showed me the opposite, the Berlin Wall has fallen. This is what makes me fight every day, fight for the rights of the people, as human beings, not as sick persons, persons with a disease, but as human beings (32 years old man with SCD, member of the Bahian Association of People with SCD)

The ABADFAL is very important, I learned a lot there... exchange of information. Today, apparently, I have a good, calm life, but there are people who have a more difficult life, you know? Then I can go there to talk or if I am in a difficulty, then a person can advise me (...) so this exchange of information I think is cool, because you bring people together. (36-years- old with SCD).

The SCD associations use many strategies to influence policymakers. Besides systematic encounters, there are national conferences where all associations gather to discuss problems and strategies. Media advocacy is also a potential tool nowadays, and all the associations communicate and share information through social media.

3.4. Biocultural Citizenship

In the beginning, there was a strong link between the Black Movement and the claims of the people with SCD, but this gradually shifted to more independent strategies. Analyzing SCD policies from 1996 until the present day leads us to interpret that the alliance with the Black Movement can be seen as what Brown et al. coined a 'window of opportunity' since racial inequities were in the political agenda and government actors were opened to address the issue [46]. It was also a very effective way of including the demands of a disease into a much broader struggle for the health of black populations, adding more advocates [47].

The social history of SCD as a disease that originated in Africa as an adaptive response to malaria evokes a very strong feeling of social belonging and blackness, which in Brazil is closely tied to racism [47]. Together with the diagnosis of SCD, a sense of 'biocultural citizenship' can be developed. Even though such an identity process was not explicitly mentioned by all study participants, this process of identity construction that claims rights to health based on ancestry was identified throughout the whole data set [48]. Here are two examples:

I think I am black, although I am fair skinned, but I am black. Because SCD is black, the carrier, appeared in Africa, about a hundred years ago, more than a hundred years ago, and came to Brazil. That is where the greatest concentration of black people and carriers are. Is it a coincidence? (...) That is also because is Brazil. If it were in Europe, they would already have it [a treatment], but because it is black blood, it is in Brazil, but if it were white blood in Europe, they would already have it. (36-years-old man with SCD)

I think that, in the case of the health policy for the black population, there are those diseases, as we already know, that affect black people the most, so I think that the government's proposal should be looking more closely at those diseases that before were not seen as much, like SCD. If it was not for this policy, it would not be today where it is, right, with all this visibility, so it was a way of putting in evidence and looking for a better quality of life for these patients. (Nurse who worked at the reference service for people with SCD at the time of the interview).

In this respect, the feeling of neglect experienced by individuals with SCD uses the existence and permanence of racism in Brazil as a means of explaining the government's disregard of SCD [47]. Therefore, racism becomes a unifier of demands and a driving force of activism for individuals with SCD [49]. The relationship between SCD and the challenges faced by the black population in Brazil is undeniable [50]. The social vulnerability experienced by the majority of the black population in Brazil intersects with the struggles involved in a chronic condition such as SCD [34], contributing to a cycle of impoverishment [49]. Either because of the caregivers who have to give up their jobs to care for these children, or because the children have to end their education early as a result of their multiple hospitalizations. Besides that, adults with SCD face difficulties in maintaining their jobs during the periodic crises caused by the disease, which can hamper social mobility and affect these families' income. These are complex factors that intersect with the institutional racism present in Brazilian society [34].

3.5. Knowledge Production Regarding SCD

As discussed in the previous sections, and illustrated with empirical data, visibility, prioritization, and action are important steps in health production, especially when it comes to neglected diseases and people [51]. Often, academic production plays an important role in producing evidence which can be translated into healthcare improvement programs and interventions. Regarding SCD, such academic efforts in producing knowledge can be seen as a strategy to overcome neglect. We need to acknowledge that 'consistent data on mortality, fatality and death rates related to SCD are not yet available for Brazil as a whole' [52] (p. 2). Epidemiological knowledge is crucial when planning health

policies, ensuring the awareness of health-related demands and guaranteeing the adequate allocation of resources in accordance with the demand. The prevalence of this disease is currently increasing worldwide, but can still be seen as an ‘orphan disease’ [45] p.599, due to the global neglect of SCD.

Partnerships between scientists and stakeholders can contribute to improving the quality of life of older adults who live with SCD, as a result of improvements in medicine and care. North–South collaborations and research partnerships could help improve treatment and reduce mortality in poorer countries [17,21]. Initiatives of international research, comparing countries with different levels of social inequalities, would help clarify the impact of social determinants on SCD outcomes. Both for the Brazilian experience in dealing with SCD combines achievements and setbacks, but would also contribute to South–South partnerships. One successful experience of such a partnership is between the governments of Brazil and Ghana, which helped to expand a national newborn screening program for SCD in Ghana [17]. It is important to keep a critical approach regarding a genetic condition that affects poorer populations worldwide, such as SCD. The global discourse around inherited blood disorders, which often uses terms such as risk, prevention, burden, etc., covers political efforts to control the population and limit their reproductive rights [53]. This discriminatory approach to SCD can contribute to stigmatizing people and legitimate eugenic policies [49,54].

4. Conclusions

At the time of writing, most of the research on SCD remains focused on clinical, pharmacological, and pathological aspects of the disease, and there continues to be a lack of qualitative research around the experiences of SCD. Although there has been a large increase in studies exploring social, socioeconomic, racial, and institutional issues in the last ten years, the number is still insufficient. It is paramount that future research on living with SCD include people who live with SCD in the planning, execution, and production of knowledge. Indeed, in one SCD meeting we heard the phrase ‘*Nothing about us, without us*’. This is a slogan frequently used by the Disability Rights Movement to communicate the idea that no policy should be decided by any representative without the full and direct participation of members of the group(s) affected by that policy. This is important too, when it comes to the process of decision making in health services, advocating for the intense and broad participation of society. In recent years, several organizations have been using this phrase as a representation of the rights of disadvantaged people, and to demand that they themselves define what their needs are and how they can be fulfilled.

Care also implies fighting racism, homophobia, and other mechanisms of social exclusion. Both healthcare managers and healthcare professionals need to reflect on the historic privileges of the white population and the historic disadvantages of the black and indigenous populations in relation to SCD. It is insufficient to recognize the health inequities that differentiate between white and black population groups in Brazil; racism must be understood as a producer and reproducer of this process of neglect. Furthermore, it is insufficient to reflect on racism as a part of the structure of capitalist society. The subjects and the institutions that drive this structure must be identified. This is an ethical and political commitment without which it will be impossible to go forward and implement truly universal and equitable healthcare for people with SCD in Brazil.

In summary, despite the advances already made in the (health)care of people with SCD in Brazil, our research and engagement reveal that there are still many challenges and obstacles to overcome. We recognize the complexity of organizing and implementing routine comprehensive SCD care. As a conclusion, we provide a set of recommendations to develop actions, drawn from our longitudinal research, for the main stakeholder groups committed to improving the lives of people living with SCD.

Recommendations for researchers

1. It is paramount that university researchers advance robust partnerships, developing and maintaining a space to listen to the priority demands of services and associations of people with SCD;
2. It is essential that academia contributes to research on the epidemiology of SCD in Brazil to comprehensively chart the impact of social determinants on health, which, in turn, will contribute to better planning of interventions;
3. In view of the advances in medicine and care, people with SCD have lived longer, making it necessary to invest in research on the health of older adults with SCD;
4. It is important that SCD is included in all curricula of health courses and that researchers contribute to the continuous development of healthcare professionals caring for people with SCD.

Recommendations the social movements involved with the SCD

1. Strengthen the associations of people with SCD, involving a wider range of advocates;
2. Be familiar with the political agendas in order to monitor and charge the inclusion of SCD and its agendas;
3. Articulate with other social movements in the health area's joint mobilization actions, as well as with activists within universities and health sectors;
4. Strengthen actions to raise awareness and popularization of SCD and to combat any type of discrimination and stigmatization of people living with this disease;
5. Increase the use of social media for communication on SCD treatments, the rights of patients and their families.

Recommendations to health policy stakeholders at national, state and municipal level

1. Support the process of producing and implementing the policy 'Sickle Cell Disease: Basic guidelines of care line' [42];
2. Activate SCD networks within each state, bringing together all social actors who work on SCR: health service providers, researchers, communities, and social movements;
3. Build and validate a matrix of indicators for monitoring and systematic evaluation of the SCD policy in each state/municipality;
4. Advance the communication between the three levels of the care network, strengthening primary care as a gateway to the system, but also ensuring access to specialized and hospital care. Strengthen primary care so that professionals can be able to perform active search of people with SCD, early diagnosis, dispensing medications, and identification of avoidable complications;
5. Ensure that the SCD-related data that are captured and available in information systems through a computerized database;
6. Advance the decentralization of haematological care, so that service users living away from urban centers can have easy access to the necessary care;
7. Advance in comprehensive care, ensuring access to a multidisciplinary network of care for people with SCD;
8. Ensure access to diagnostic and follow-up tests and the uninterrupted supply of medicines for people with SCD.

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