

RESEARCH ARTICLE

# Health Care for People with Sickle Cell Disease in a Medium-Sized Brazilian City

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## Abstract:

The objective of this study was to evaluate the access, assistance and satisfaction of people with sickle cell disease in relation to the health care provided by the Unified Health System in Uberlândia city, Minas Gerais, Brazil. Thirty-four people with sickle cell disease were recruited using the snowball sampling method and submitted to semi-structured interview in which demographic, clinical, relationship and satisfaction with health care were collected. Forty-four percent were not included in the Primary Health Care network, which did not provide several procedures/actions recommended by the Ministry of Health. Ninety-four percent were unsatisfied/partially satisfied with the emergency care provided in the Integrated Care Units. The main reasons for the unsatisfied were ignorance of sickle cell disease by the health team (87.5%), delay in care (81.3%) and inadequate conduct by the health team (59.5%). In conclusion, the access and quality of care provided to people with sickle cell disease by the Primary Health Care network and the Urgency and Emergency Network need to be improved and this improvement is directly related to the training of health teams.

**Keywords:** Sickle cell disease, Primary health care, Evaluation of health care.

## Introduction

Sickle cell disease (SCD) is one of the most prevalent monogenic hereditary diseases in Brazil and worldwide, being considered an important public health

problem [1-3]. Originally from Africa, it was brought to Brazil by the forced migration of slaves, most often affecting black and brown people. According to the National Neonatal Screening Program, it's estimated that 3,500 children with SCD and 200,000 with sickle cell trait are born in Brazil each year [4,5].

Transmitted in an autosomal recessive pattern, SCD results from a mutation in the beta-globin gene, with the formation of an anomalous hemoglobin called hemoglobin S (HbS). In addition, SCD encompasses a group of hemoglobinopathies in which the HbS gene is present in homozygosis (SS genotype) or in compound heterozygosis that is combined with another hemoglobin variant such as hemoglobin C (SC genotype),  $\beta$ -thalassemia (S $\beta$  genotypes), hemoglobin E (SE genotype) and others rarer. The SS genotype has a more severe clinical significance, being called sickle cell anemia (SCA) [6-8].

The HbS has altered physical and chemical properties, showing a propensity to undergo polymerization under conditions of low oxygen tension, which leads to alteration of red blood cell shape (sickle shape). Sickle red blood cells present several changes in their membrane, becoming rigid, more mechanically fragile, therefore, more susceptible to intravascular hemolysis and with increased adhesion to endothelial cells, leukocytes and platelets. Thus, they are easily trapped in places with slower microcirculation, generating acute vasoocclusive events with ischemic and reperfusion injuries in various organs and tissues [9,10].

The most relevant clinical events of the disease are acute and recurrent crises of pain, chronic hemolytic anemia, progressive impairment and insufficiency of multiple organs. These manifestations, despite varying from individual to individual, are potentially devastating and, in general, have important repercussions in various aspects of a person's life, it may even reduce life expectancy [11-14]. Thus, as a chronic disease, which persists throughout life, usually incurable, but treatable, SCD causes its carrier to require careful and continuous assistance by a multidisciplinary health team. This assistance, when properly performed, promotes morbidity and mortality reduction with an improvement in life's quality and an increase in life expectancy [15-17].

In order to ensure that person with SCD has adequate access, reception and assistance in health services that are part of the Unified Health System (UHS), the Ministry of Health of Brazil published several ordinances that defined the guidelines of the National Policy for Comprehensive Care for People with Sickle Cell Disease [18]. These guidelines include diagnostic, prophylactic, propaedeutic and therapeutic actions. It was postulated that Primary Health Care Network (PHCN) should be the preferred gateway for people with SCD in to the health system, with Family Health Strategy (FHS) team as the organizing center for all services in all assistance levels (low, medium and high complexity). Basic Health Care (BHC) units must be integrated with specialized care reference centers and coordinate the necessary care. Various clinical protocols and therapeutic guidelines were also developed to qualify this care and it was determined that, at all levels of care, the qualification and continuing education of health professionals involved in this care should be promoted [19-22].

This study aimed to assess the access, assistance and satisfaction of people with SCD relation to health care provided by the UHS network in the Uberlândia city, Minas Gerais, Brazil.

## Materials and methods

### *Study location*

This is a descriptive, cross-sectional, quali-quantitative study in which people with SCD who lived in the Uberlândia city participated.

Uberlândia city is located in the southeast of Brazil being the second largest city in population in Minas Gerais state, estimated 691,305 inhabitants by the Brazilian Institute of Geography and Statistics in 2019 [23]. According to a report made available by the website of the National Register of Health Establishments [24], the Public Health network of Uberlândia city, that provides services to UHS, has 56 Family Health Teams (FHT), 12 conventional Basic Health Units (BHU), 8 Integrated Care Units (ICU), an university hospital (belonging to the Federal University of Uberlândia) and a Blood Center (belonging to the Hemominas Foundation), providing assistance at the level of primary care and complexity medium and high.

### *Participants recruitment*

Participants were recruited using the snowball sampling technique [25] in which, briefly, a key informant, known to have SCD, was found. This person was asked to indicate the contact of other people who also had the disease. The process continued until the minimum sample size was reached and slightly exceeded.

### *Sample size determination*

The methodology proposed by Fonseca & Martins was used to sample size determination [26]. Knowing that Uberlândia city population in the age group between 18 and 54 years old is approximately 400,000 people [23], that the incidence rate of SCD in Minas Gerais state is around 1:1400 live births [27] and that a sampling precision of 1% around the central value and a 95% confidence level were desired, it was calculated that the minimum sample size should be 27 participants.

### *Participants*

Thirty-four people with SCD participated in the study, who met the inclusion criteria: being 18 years of age or older (laboral age), living in Uberlândia city and not having cognitive impairment or having other relevant chronic diseases not related to SCD. All of them had a SCD diagnosis confirmed by the hemoglobin electrophoresis test, performed previously at some time in their lives, whose result (genotype) was in the data in the Blood Center's registry. After signing a free and informed consent form, they were submitted to a semi-structured interview, with collection of demographic, clinical, relationship and satisfaction data with healthcare. The interviews were conducted from November 2018 to January 2019.

### *Analysis*

The data obtained were recorded in a Microsoft Excel electronic data base and submitted to an exploratory descriptive statistical analysis (frequencies and percentages for categorical variables and measures of central tendency and dispersion for continuous variables). All analyzes were performed using SPSS 20 software.

### *Ethics committee*

All experimental procedures were approved by Research Ethics Committee (CEP) of the Federal University of Uberlândia, with protocol number 2.985.296.

## Results

The thirty-four participants were aged between 18 to 70 years old, with an average age of  $35.82 \pm 15.29$  years old. Only one (2.9%) was 70 years old and about 65% was 40 years old or younger. Regarding gender, 55,9% of the sample was male. The majority (79.4%) had black or brown skin (Table 1). Eighteen individuals (53.9%) had completed high school. Of these, five (14.7%) were in higher education and one (2.9%) had already completed it. Eleven people (32.4%) had only elementary education and of these, seven (20.6%) did not complete it.

The most common hemoglobin genotype was SS (64.7%) and eight people (23.5%) had their diagnosis established by neonatal screening (Heel Prick Test). Thirty-three (97.1%) had already been hospitalized due to complications related to SCD. Fifteen participants (44.1%) used hydroxyurea (Table 1).

Table 1: Demographic characteristics of the participants and use of hydroxyurea in relation to the disease genotype.

Characteristics of the participants	Genotype			
	SS (N=22) 64,7%	SC (N=11) 32,4%	S $\beta$ (N= 1) 2,9%	Total (N=34)
Sex				
Female	5 (33,3)	9 (60,0)	1 (6,7)	15 (44,1)
Male	17 (89,5)	2 (10,5)	0 (0)	19 (55,9)
Skin				
Black	11 (57,9)	8 (42,1)	0 (0)	19 (55,9)
Brown	7 (87,5)	1 (12,5)	0 (0)	8 (23,5)
White	4 (57,1)	2 (28,6)	1 (14,3)	7 (20,6)
Age group (years)				
18 - 21	6 (60,0)	4 (40,0)	0(0)	10 (29,4)
22 - 49	14 (82,4)	3 (17,6)	0 (0)	17 (50,0)
50 - 70	2 (28,6)	4 (57,1)	1 (14,3)	7 (20,6)
Hydroxyurea				
No	5 (35,7)	8 (57,2)	1 (7,1)	14 (41,2)
Yes	12 (80,0)	3 (20,0)	0 (0)	15 (44,1)
Already used in the past	5 (100,0)	0 (0)	0 (0)	5 (14,7)

Regarding access and use of health system, 15 participants (44.1%) weren't included in Primary Health Care network as a result of living in areas without coverage. This contrasted with the monitoring at the Specialized Service, carried out by the Regional Blood Center of Uberlândia, in which 32 people (94.1%) were followed up. The 19 participants (55.9%) inserted in the BHC were questioned regarding the availability by this network of the procedures and actions recommended by the Ministry of Health (Table 2).

Table 2: Availability in the Basic Health Units (BHU) and Basic Family Health Units (BFHU) of the procedures/actions recommended by the Ministry of Health.

Procedures/actions	Availability in health units (BHC and BFHU)			Total=19
	No N (%)	Yes N (%)	Do not know N (%)	
Electrophoresis Hb	9 (47,4)	0 (0)	10 (52,6)	
Sickle cell trait orientation	17 (89,4)	1 (5,3)	1 (5,3)	
Forwarding of exams requested by the specialized service	4 (21,1)	15 (78,9)*	0 (0)	
Supply of medications protocol	16 (84,2)	3 (15,8)**	0 (0)	
Supply of vaccines protocol	0 (0)	19 (100)	0 (0)	
Wound prevention and forwarding to a referral center	12 (63,2)	0 (0)	7 (36,8)	
Link maintenance	17 (89,5)	2 (10,5)	0 (0)	
Identification and forwarding of urgencies/emergencies	19 (100)	0 (0)	0 (0)	
Promotion of social inclusion	17 (89,5)	0 (0)	2 (10,5)	

\* About 80% of patients who stated that BHC conducts the examinations requested by the Specialized Service also reported that they enter a waiting list with an average delay of 2 years for the examinations.

\*\* The units' pharmacies only supply medicines when they are made available by the municipality, which is not always the case.

Emergency care is provided by ICU and when asked about satisfaction with this service, only two participants (5.9%) were satisfied, 20 (58.8%) were unsatisfied and 12 (35.3%) partially satisfied (Table 3). The main reasons, spontaneously reported, for this unsatisfied/partial satisfaction were, in decreasing order, ignorance of SCD by the health team, delay in service and inadequate conduct by the service team (Table 4).

Table 3: Satisfaction of study participants with urgent/emergency care in Integrated Care Units (ICU) and Specialized Service (Hemocentro).

Service	Satisfaction			N=34
	Unsatisfied N (%)	Satisfied N (%)	Partially satisfied N (%)	
Integrated Care Units (ICU)	20 (58,8)	2 (5,9)	12 (35,3)	
	Unsatisfied N (%)	Satisfied N (%)	Partially satisfied N (%)	N=32
Specialized Service (HEMOCENTRO)	0 (0)	24 (75,0)	8 (25,0)	

Table 4: Main reasons for unsatisfied/partially satisfied of 32 study participants in relation to emergency care in Integrated Care Units (ICU).

Reasons for unsatisfied/partially satisfied	N (%)
Not knowledge of sickle cell disease by the health team	28 (87,5)
Service delay	26 (81,3)
Inappropriate conduct by the health team	19 (59,5)
Lack of unit structure	15 (46,9)

Between 32 participants accompanied by the Specialized Service, 24 (75%) are satisfied with this service and eight (25%) partially satisfied (Table 3). The partially satisfied ones complained of difficulty in understanding the language used in the orientation (33.3%), unpreparedness of the physiotherapy team (22.2%) and precarious resources for dental treatment (11.1%) (Table 5).

Table 5: Main reasons for the partial satisfaction of eight study participants in relation to attendance at the Specialized Service (Hemocentro).

Reasons for partially satisfied	N (%)
Difficulty understanding the language used in orientation	26 (33,3)
Lack of preparation of the physiotherapy team	15 (22)
Precarious resources for dental treatment	19 (59,5)

## Discussion

Our study included only people with SCD who were at least 18 years old and didn't have cognitive deficits. These two criteria associated with the fact that most of them (53.9%) had completed high school suggest that the reports and opinions they issue are consistent with the realities experienced.

The slight predominance of the male gender was also observed in Fernandes's study and colleagues [28], in which 52.8% of children hospitalized with SCD were of the same gender. On the other hand, some studies show higher prevalence of females [29,30], demonstrating that SCD is a genetic disease not linked to sex.

The highest frequency of the SS genotype (sickle cell anemia) among the participants is in line with the fact that it's the most frequent worldwide [7]. Similarly, confirming what is already observed worldwide, a greater number of blacks and browns people affected by the disease were also found. This finding is relevant to the extent that this population group has, in general, the worst epidemiological, educational and economic indicators, which contributes to a worse prognosis of the disease [11,31].

Interestingly, seven people (20.6%) aged between 50 and 70 years were found and among them, three were considered elderly (aged 60 or more). This is a curious fact because it's an unusual finding in SCD.

In a study that evaluated 3,764 people with SCD, that the median age at the time of death for patients with the SS genotype was 42 years for men and 48 years for women and 60 years for men and 68 years for women with SC genotype [32]. These data corroborate our findings, since the three elderly individuals had the SC genotype.

All participants, except one, had already been hospitalized. This single exception corresponded to the 70 year old individual with the SC genotype and can be explained by the clinical variability of the disease. This variability can be associated with genetic factors, such as genotype and environmental factors, such as socioeconomic status, place of residence, access and quality of medical care and prevalence of infectious diseases [15].

In Minas Gerais state, the State Neonatal Screening Program (Heel Prick Test) implemented screening for hemoglobinopathies in March 1998, so it has been 21 years in relation to this study. In recent years, 98% of newborns in Minas Gerais state have neonatal screening. The objective of this program, in addition to the early diagnosis of SCD, the institution of adequate treatment and prophylactic measures that contribute to a significant morbidity and mortality reduction [33]. In our study, 75% of participants aged between 18 to 21 years old had their diagnosis confirmed by this procedure, reflecting the initial results of implementing the program in Uberlândia city.

Hydroxyurea is a chemotherapeutic agent, myelosuppressant, it's currently the only drug with proven efficacy in the treatment of SCD. Some of the benefits resulting from the hydroxyurea use in SCD are reduced frequency of painful episodes, reduced transfusion needs and episodes of acute chest syndrome, increased production of fetal hemoglobin (HbF) and decreased risk of death. Possible adverse effects from its use include neutropenia, bone marrow suppression, elevated liver enzymes, anorexia, nausea, vomiting and infertility [34,35].

In our study, about 44% of participants were using hydroxyurea. Between these participants, 80% had the SS genotype and the others the SC genotype. This finding is justified since the clinical severity of SCD in patients with the SS genotype (sickle cell anemia) is generally greater [15,36] and the prescription of hydroxyurea follow the mandatory presence of some inclusion criteria. Among these is the occurrence, in the last 12 months, of at least one of a list of complications such as: three or more acute pain episodes requiring medical attention, more than one event of acute chest syndrome, chronic hypoxemia, chronic organ injury proven, Hb < 7g/dL out of acute event, HbF concentration < 8% after 2 years old, leukocyte count > 20,000/mm<sup>3</sup>, lactic dehydrogenase levels twice above the reference value for age and changes in transcranial echocardiography [37]. This means that at least 44% of the survey participants had at least one of these complications.

Accessibility to health care can be understood as the ability to generate services and meet the health demands for a given group, including the characteristics of services and resources that facilitate or prevent their use by users. Therefore, access to a health service or service network is influenced by multiple factors such as geographic, organizational, socio-cultural and economic [38].

In the present study it was observed that the access of people with SCD to the Primary Health Care network in Uberlândia city is seriously compromised, since a significant portion of the study participants (44.1%) is not included in this network in due, among other reasons, to living in places without coverage. On the other hand, we found that 94.1% of the studied population is served by the Specialized Service, represented by the Blood Center, regardless of whether they live at a reasonable distance from this institution. This is nothing new, repeating what was observed in other studies carried out in various parts of the country. One of the reasons postulated for this is that historically the treatment of SCD is considered as the competence of hematological centers [11,39].

Perceptibly, there are other reasons for this to happen, which is evident when we analyze satisfaction with the services provided by BHC and the Blood Center. The number of unsatisfied/partially satisfied with the assistance provided by BHC was very expressive (94.1%), in contrast to the also expressive satisfaction (75.0%) shown in relation to the Blood Center. Spontaneous reporting of reasons for unsatisfied/partial satisfaction allows for a better understanding of the care situation as a whole. The most frequent complaints, in relation specifically to the emergency care provided by ICU, and the reports in the literature refer us to some possible basic reasons for these occurrences: failure in the humanization process of service and unpreparedness of the health teams to care for the person with SCD. This unpreparedness is undoubtedly result of training lack of health teams, which perpetuates the status of ignorance the disease and significantly limits the possibility of providing comprehensive and adequate assistance to people with SCD.

The manual “Sickle Cell Disease - Basic Guidelines for the Care Line”, prepared by Ministry of Health [18], postulates that humanization must be based on providing a comfortable environment for care of people. These must be received in a caring and supportive manner, without showing prejudices and stigmas; they must be attentively listened, communication must be respectful and accessible language, easy to understand to the lay public should be used.

Classically, people with SCD have recurrent and disabling pain episodes. Worldwide there are numerous publications with reports of stigmatization of adults with SCD in health system, such as health professionals whose generally insensitive to their experience of pain. People with SCD are often seen by the health team as drug addicts. These negative experiences with health care make many of these people delay the search for medical care managing their episodes of pain at home. Therefore, having prior guidance to be able to perceive and understand the pain experience of the person with SCD and to properly welcome them is the first step to improve access and assistance to this population [13,40].

The second most frequent complaint of the study participants refers to the delay in attending when they sought the ICU emergency service. The investigation and recognition of warning signs in patients with SCD make their care a priority. This suggests that health teams are not prepared to recognize the signs and needs in care of people with SCD [38,39,41]. It's noteworthy that participants themselves detected the lack of knowledge of the health team. This

was the main reason cited by them for their dissatisfaction with emergency care and inadequate treatment.

As previously mentioned, economic and organizational factors also influence the access and efficiency of a health service [39]. Perhaps these two factors were preponderant in determining the findings regarding the availability of procedures/actions (recommended by the Ministry of Health) in Basic Health Unit and Basic Family Health Unit (BFHU). Procedures/actions such as: informing the user that the network provides hemoglobin electrophoresis test for relatives and partner, promoting the maintenance of the bond through regular visits, providing guidance on the sickle cell trait and the disease, performing the preventing wounds, identifying and forwarding urgencies/emergencies, which are not being carried out or carried out minimally, probably depend more on organizational procedures and training of the health team than economic factors. These procedures/actions are relevant, as they change attitudes and decrease physical, emotional and social problems resulting from the disease [15,39,42].

A limitation of our study is due to the sampling method used (snowball sampling method), which may have led to the constitution of a sample that is not so representative of the population of people with SCD in Uberlândia city.

## Conclusions

The unsatisfied shown by SCD patients in relation to care provided by Public Health System demonstrates important deficiencies in management and provision services by health system and need for both access and quality of care to be improved. This unsatisfied was greater in assistance provided by Primary Health Care and in attendance to emergencies/urgencies, practically not being verified in relation to specialized service linked to Hemocentro, which for this reason is the most sought by patients.

The lack of knowledge about the disease, the unpreparedness and the inadequate conduct of health teams, as well as the failures in reception, identified by SCD patients as relevant to their unsatisfied, demand urgent and effective interventions. It is essential to implement organizational and training procedures of health teams of BHC, FHT and ICU which guarantee qualified assistance to SCD patients allowing a better quality of life and avoiding suffering, serious and disabling sequelae.

In conclusion, the performance of public health managers, municipal and state, who have knowledge of situation, promote the training of health professionals and their humanized acting, use mechanisms to monitor the performance and efficiency these professionals and create strategies to provide in health services actions and resources essential recommended by Ministry of Health guidelines.

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