Impact of sickle cell disease on work activity
Impacto da doença falciforme na atividade laboral

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ABSTRACT | Introduction: Sickle cell disease is a group of conditions caused by anomalous hemoglobin that predisposes people to some clinical syndromes. Because of these recurrent syndromes, patients have difficulty finding and, often, keeping a job. Objectives: To evaluate the impact of sickle cell disease on the job situation of people with the condition. Methods: Thirty-two working-age people with sickle cell disease were recruited using the snowball sampling method and underwent semistructured interviews for demographic, socioeconomic, and clinical data collection. Results: Only 28.1% of the interviewees engaged in paid work, 46.9% had already worked but were not working at the time of the interview, and 25% had never worked. About 6% of participants lived in extreme poverty, and 28.4% lived on the poverty line. Monthly per capita income was less than one minimum salary in 56.2% of cases and less than 1.5 minimum salaries in 9.4%. Conclusions: Sickle cell disease has an important negative impact on employment situation, as about 70% of working-age people were inactive. This results in a high social cost represented by a very low monthly per capita income (≤ 1 minimum salary) in 93.7% of the participants. Keywords | sickle cell disease; work performance; monthly per capita income.

RESUMO | Introdução: A doença falciforme é um conjunto de enfermidades ocasionadas pela hemoglobina anômala que predispõe a algumas síndromes clínicas. Devido a essas síndromes recorrentes, os portadores dessa doença têm dificuldade em encontrar e, muitas vezes, permanecer no emprego. Objetivos: Avaliar o impacto da doença falciforme na situação laboral de pessoas portadoras da doença. Métodos: Trinta e duas pessoas com doença falciforme, em idade laboral, foram recrutadas utilizando-se o método “bola de neve” e responderam a uma entrevista semiestruturada para coleta de dados demográficos, socioeconômicos e clínicos. Resultados: Apenas 28,1% dos entrevistados estavam exercendo uma atividade remunerada, 46,9% já tinham trabalhado, mas não trabalhavam mais à época da entrevista e 25% nunca tinham trabalhado. Cerca de 6% dos participantes viviam em extrema pobreza e 28,4% viviam na linha da pobreza, sendo a renda mensal per capita menor que um salário mínimo em 56,2% dos casos e menor que 1,5 salário mínimo em 9,4%. Conclusões: Concluímos que a doença falciforme exerce importante impacto negativo na situação laboral, com cerca de 70% das pessoas em idade laboral sendo inativas. Isso resulta em um alto custo social, representado por uma renda mensal per capita muito baixa (≤ 1 salário mínimo) de 93,7% dos participantes. Palavras-chave | doença falciforme; atividade laboral; renda mensal per capita.
INTRODUCTION

Sickle cell disease (SCD) designates a set of hereditary hemolytic anemias characterized by a genetic alteration that causes a hemoglobin mutation called hemoglobin S (HbS) instead of normal hemoglobin, called hemoglobin A (HbA). Among the several subtypes of hemoglobinopathies in SCD, the most severe clinical form is sickle cell anemia, which is determined by the presence of homozygous HbS (HbSS). In other subtypes, HbS is combined with other variant hemoglobins such as hemoglobin C (SC disease) and β thalassemia (S/β thalassemia disease).

SCD is one of the most common hereditary diseases in the world. In Brazil, HbS was introduced by the forced migration of Black Africans into slave labor. SCD is heterogeneously distributed, being more frequent in populations of afro-descendants. In Minas Gerais state, the prevalence is relatively significant: the homozygous form (HbSS) is found in the proportion of 40 cases/100,000 population and the heterozygous form (HbSC), 31 cases/100,000 population. Severe forms have a significant prevalence in Minas Gerais. Thus, they are recognized as an important public health problem with a great impact on the morbidity and mortality of the affected population, accounting for approximately 3.4% of total deaths in children under 5 years old. As a result, a free neonatal screening program was implemented in Minas Gerais in March 1998, as part of the heel prick screening test, with the aims of promoting prophylactic treatment and reducing morbidity and mortality from the disease. According to data from the Brazilian National Neonatal Screening Program, three thousand new cases of SCD are estimated to appear each year as well as 180 thousand new carriers of the sickle cell trait.

Work activities can be considered the main temporal organizer of life. It is through work that individuals interact and transform the environment, ensure survival, and establish interpersonal relationships, which theoretically serve to reinforce their identities, contributing to insertion in society and preservation of health.

Chronic diseases have a great impact on work performance, causing changes and interruptions in the work activity together with impaired productivity and even job loss, factors that trigger socioeconomic, emotional, and psychological problems. This category includes SCD. By determining symptoms of recurrent pain and complications that affect almost all organs and that can be exacerbated or precipitated by environmental factors, SCD has a profound impact on the ability to initiate, perform, and maintain a work activity, limiting the job options. Also, aggravating factors include low educational level and inadequate or deficient care provided by the health services.

The historical invisibility of SCD in Brazil determined by the lack of knowledge of the disease by health professionals and public managers produces inadequate assistance provided by the health system, which leads to misdiagnosis of complications, inadequate treatments, and long searches for resolutive care in several health units, which affects even further the individual’s work situation. Most probably, people with SCD have either never been employed or lost their jobs because of problems related to the disease or are underpaid. Thus, in general, their only option is the informal sector. Because of their nonprofessional status, they are subjected to jobs that require great physical effort with high oxygen consumption, which contributes to a worsening health status. The insecurity and fear determined by these situations make them uncompetitive in the labor market.

Therefore, this study aimed to evaluate the impact of SCD on the employment situation of people with the condition in Uberlândia, Minas Gerais state.

METHODS

This descriptive cross-sectional study was carried out from November 2018 to January 2019, and recruitment was based on the snowball sampling method. We selected for the study working-age (18 to 65 years) people with SCD living in Uberlândia with no cognitive impairment and no other relevant
chronic diseases not related to SCD. All experimental procedures were approved by the research ethics committee of the Universidade Federal de Uberlândia with protocol number 2.985.296.

The sample size was calculated according to a method proposed by Fonseca & Martins with an established minimum of 27 participants. We considered that Uberlândia’s population in the range of 18-54 years old is approximately 400,000 and that the incidence of SCD in Minas Gerais is nearly 1:1400 live births, with an expected sampling precision of 1% around the central value and a 95% confidence interval.

Thirty-two people met the inclusion criteria and participated in the study. Nine (28.1%) participants were currently working, eight (25%) had never worked, and 15 (46.9%) had previously worked but were not working at the time of the interview. Regarding the participants who had already worked and were not working anymore, the length of stay in the job ranged from 3 to 240 months (20 years). The age at the initiation of the work activity ranged from 12 to 38 years.

All participants were asked to sign an informed consent form and then underwent a semistructured interview for obtaining demographic, socioeconomic, and clinical data.

Statistical analysis was performed using SPSS 2.0 software. Quantitative variables were described by means and standard deviations, and qualitative variables were described by frequencies and percentages. Nonparametric Mann-Whitney U test was used to compare two independent samples, and Fisher exact test was used to compare two samples with binomial variables.

RESULTS

Age ranged from 18 to 60 years with a mean of 33.78 ± 13.04 years. Fifty-nine percent of the participants were male, and 68.4% were currently inactive. Twenty-five (78.1%) participants stated that they had Black/Brown skin color, and within this group, 18 (72%) were inactive. Twenty-four (68.8%) participants had the SS genotype, characteristic of sickle cell anemia, and 15 (68.2%) were inactive (Table 1).

The most frequent level of education among the participants was high school, as 53.1% had completed

| Table 1. Demographic characteristics, schooling, genotype, diagnosis by the heel prick screening test, and use of hydroxyurea in relation the work situation of the participants |
|-----------------------------------------------|
| **Features** | Total n (%) | Work situation | | |
| | | Active n (%) | Inactive n (%) | p-value |
| Sex | | | | |
| Female | 13 (40.6) | 3 (23.1) | 10 (76.9) | 0.70* |
| Male | 19 (59.4) | 6 (31.6) | 13 (68.4) | |
| Skin color | | | | |
| Black/Brown | 25 (78.1) | 7 (28.0) | 18 (72.0) | 1.00* |
| White | 7 (21.9) | 2 (28.6) | 5 (71.4) | |
| Age (years) | | | | |
| 18-21 | 10 (31.3) | 3 (30.0) | 7 (70.0) | 0.47* |
| 22-49 | 17 (53.1) | 6 (35.3) | 11 (64.7) | |
| 50-60 | 5 (15.6) | 0 (0.0) | 5 (100.0) | |

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high school. One participant (3.1%) had already completed higher education and five (15.6%) had not yet completed higher education. Despite having a complete educational level, 18 (43.75%) participants were inactive. Regarding the use of hydroxyurea, 20 (62.5%) participants had already been medicated and 12 (37.5%) participants had never taken this medication (Table 1).

Monthly per capita income was less than or equal to one minimum salary in 29 (90.55%) participants, with a minimum amount of BRL 90.00 and a maximum amount of BRL 1,497.00 (Table 2).

### Table 1. Continued

| Features                        | Total n (%) | Work situation | p-value |
|---------------------------------|-------------|----------------|---------|
|                                |             | Active n (%)   | Inactive n (%) |
| Educational level               |             |                |          |
| Incomplete elementary school    | 6 (18.8)    | 2 (33.3)       | 4 (66.7)  | 0.35† |
| Complete elementary school      | 4 (12.5)    | 1 (25.0)       | 3 (75.0)  |      |
| Incomplete high school          | 5 (15.6)    | 3 (60.0)       | 2 (40.0)  |      |
| Complete high school            | 11 (34.4)   | 2 (18.2)       | 9 (81.8)  |      |
| Incomplete higher education     | 5 (15.6)    | 1 (20.0)       | 4 (80.0)  |      |
| Complete higher education       | 1 (3.1)     | 0 (0.0)        | 1 (100.0) |      |
| Hydroxyurea use                 |             |                |          |
| Never                           | 12 (37.5)   | 2 (16.7)       | 10 (83.3) | 0.66† |
| Previously                      | 5 (15.6)    | 1 (20.0)       | 4 (80.0)  |      |
| Currently                       | 15 (46.9)   | 4 (26.7)       | 11 (73.3) |      |
| Hemoglobin genotype             |             |                |          |
| SS                              | 22 (68.8)   | 7 (31.8)       | 15 (68.2) | 0.80† |
| SC                              | 9 (28.1)    | 2 (22.2)       | 7 (77.8)  |      |
| Sβ                              | 1 (3.1)     | 0 (0.0)        | 1 (100.0) |      |
| Heel prick screening diagnosis  |             |                |          |
| No                              | 24 (76.5)   | 5 (20.8)       | 19 (79.2) | 0.18* |
| Yes                             | 8 (23.5)    | 4 (50.0)       | 4 (50.0)  |      |

SC = SC hemoglobin  
SS = SS hemoglobin  
Sβ = Sβ hemoglobin  
* Fisher exact test for two samples with binomial variables.  
† Mann-Whitney U test for two independent samples.

### Table 2. Distribution of monthly per capita income

| Monthly per capita income (BRL) | Participants n (%) |
|---------------------------------|--------------------|
| 9000-14500                      | 2 (6.25)           |
| 14600-42000                     | 9 (28.10)          |
| 42100-99800*                    | 18 (56.20)         |
| 999900-149700†                  | 3 (9.40)           |

Base salary for the year 2019. Additionally, 12 (37.5%) participants lived in rented houses, 2 (6.3%) in houses provided by friends, 8 (25%) in financed houses, 1 (3.1%) in an appropriation area, and only 9 (28.1%) in their own houses (data not shown).  
†1 minimum salary.  
†1.5 minimum salaries.

### DISCUSSION

Work activity and chronic disease have an important mutual impact. As a determinant of socioeconomic level, a work activity influences the status and prognosis of a disease with an impact on variables such as access to medical care, rapid treatment of crises, adequate basic sanitation with reduced risk of infection, high-quality nutrition, and better living conditions. Additionally, diseases...
significantly influence the performance of a work activity.\textsuperscript{9}

To assess the impact of SCD on work activity, 32 working-age people with SCD were interviewed. Although most participants were under 40 years old, 11 people were 40 years old or over, and 5 of those were between 50 and 60 years old. Given that SCD is characterized by a reduced life expectancy with early mortality,\textsuperscript{15} those findings are interesting. Data released by the Brazilian Ministry of Health\textsuperscript{16} referring to a North American study showed a life expectancy of 42 years old for men and 48 years old for women with SCD. This seems to show a tendency to increased life expectancy in the population of interest, although any assumption in that sense is still very early and involves no solid reasoning.\textsuperscript{17}

Regarding sex and skin color, we observed a slight predominance of men (SCD is an inherited disease not linked to sex) and a strong predominance of Black/Brown skin color (nearly 80% participants). The predominance of Black skin color can be understood by the specificity of the disease, as SCD is directly related to the Black population.\textsuperscript{18} These results have some relevant implications. For example, according to data from the Brazilian Institute of Geography and Statistics (Instituto Brasileiro de Geografia e Estatística, IBGE), 72.7% of people living below the poverty line are Brown and Black.\textsuperscript{19} In addition, this population group suffers from worse economic, epidemiological, and educational indicators, which contributes to a poorer prognosis of SCD with reflections on work capacity.\textsuperscript{9,20}

Our study showed that SCD is an important determinant of impaired work capacity, as only nine (28.1%) participants were active. The impact of that on the lives of people with SCD should not be underestimated. Work is an essential activity not only for livelihood but also for the maintenance of physical and mental well-being, being perceived as something that gives meaning to life and drives human growth.\textsuperscript{21,22} According to the Brazilian Ministry of Health in the Workers’ Guide, work or absence from work determines health situation, enables material condition, and promotes social inclusion.\textsuperscript{23}

There is a relationship between diseases, disabilities, and restrictions according to Swanson et al.\textsuperscript{24} In their study, SCD was combined with the following terms: pain, employment, vision, hearing, cognition, school performance, language, depression, mobility, and emotional, educational, and behavioral problems. The authors found that adults with SCD had high levels of unemployment and nonparticipation in the workforce, loss of workdays due to painful crises, and unfavorable attitudes of their employers.\textsuperscript{24}

Paiva e Silva et al.\textsuperscript{20} reported that difficulty finding a job and an insufficient financial situation are among the main causes of the onset of depressive status in patients with SCD. They reached this conclusion by studying 80 people with SCD and observing that socioeconomic level was extremely low with a \textit{per capita} income below one and a half minimum salary in 85% of the cases.\textsuperscript{20}

We were unable to demonstrate, probably because of the small sample size (32 participants), a statistically significant relationship between employment status and demographic characteristics, education, use of hydroxyurea, mode of diagnosis, and disease genotype.

Regarding the use of hydroxyurea, Ballas et al.\textsuperscript{25} compared the employment status of people with SCD who used the drug and responded to treatment and others receiving placebo. They observed that hydroxyurea treatment did not significantly affect employment status; however, there was a trend towards more consistent employment in the hydroxyurea group. Nonetheless, they stressed the importance of treating young patients with hydroxyurea to prevent or mitigate the incidence of complications and thus improve future employment status.\textsuperscript{25}

All participants in our study had an extremely low socioeconomic status. Two participants (6.25%) and their families survived on a monthly \textit{per capita} income of less than BRL 145.00, which is considered living below the extreme poverty line. Nine participants (26.47%) and their families survived on less than BRL 420.00 monthly \textit{per capita} income; therefore, they lived on the poverty line. Twenty-nine participants (90.6%) received up to BRL 954.00 per month, which is less than the minimum salary in 2019 (BRL 998.00). This percentage was higher
than that found by Felix et al., who analyzed the monthly income of 47 people with SCD and observed that 48.9% received up to a minimum monthly salary.

According to the IBGE Social Indicator Synthesis, a quarter of the Brazilian population lives on less than BRL 420.00 monthly per capita income. According to 2019 data from IBGE, 72.7% of people living below the poverty line are Brown and Black, a group that represents the majority of the participants in our study.

Regarding housing, we observed that only nine (28.1%) participants had their own houses. We could not find in the literature information related to housing and SCD. However, when considering the Minha Casa, Minha Vida Program, a housing program targeting people living on a minimum income whose priority criterion is the presence of a chronic disease, we believe that this program may have contributed to more people owning a house. Nonetheless, we can infer that public policies do not yet effectively and comprehensively address people with chronic diseases, who require special care and attention from the government.

Twelve (37.5%) participants lived in rented houses, two (6.3%) in houses provided by friends, eight (25%) in financed houses, one (3.1%) in an appropriation area, and only nine (28.1%) in their own houses.

**CONCLUSIONS**

SCD has an important negative impact on work activity, leading to a significantly impaired per capita and family income for people with the condition and contributing to a reduced quality of life. Measures to minimize this impact are urgently needed and encompass two action fronts. The first refers to a health promotion strategy aiming to ensure that people with SCD have adequate access to primary care and education services in an attempt to promote self-knowledge and self-care. The second involves the implementation of policies that protect people regarding job admission and working conditions and that encourage employers to hire and keep them.

**Author contributions**

RPP, MCO, LBA, JCO, and TMA were responsible for the study conceptualization, formal analysis, and writing – review & editing of the text. RPP participated in the investigation and funding acquisition. RPP, MCO, and LBA participated in data curation. RPP, JCO, and TMA participated in project administration and methodology. JCO and TMA participated in the supervision and validation of the study. All authors approved the final version submitted and assume public responsibility for all aspects of the work.

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