HEMATOLOGY, TRANSFUSION AND CELL THERAPY

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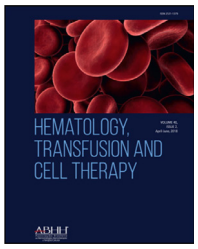

Original article

Prejudice impairing quality of life in sickle cell disease patients in a developing country: faces of suffering

Camila Freitas de Andrade Rodrigues *, *Thiago Alves Rodrigues*, *Eduardo José Silva Gomes de Oliveira*, *João Batista Santos Garcia*, *Maria do Socorro de Sousa Cartágenes*

Centro de Ciências Biológicas e da Saúde, Universidade Federal Universal do Maranhão (CCBS UFMA), São Luís, MA, Brazil

ARTICLE INFO

Article history:
Received 30 March 2021
Accepted 13 June 2021
Available online 13 July 2021

Keywords:
Sickle cell disease
Prejudice
Quality of life

ABSTRACT

Introduction: The perception of prejudice against, and stigmatization of, sickle cell disease (SCD) leads the patient to perceive a different treatment, due to the disease stigma and may be related to a worse quality of life (QoL).
Objectives: Describe and evaluate the perception of the prejudice against the disease and its impact on the quality of life of patients with sickle cell disease.
Methods: This is a cross-sectional study conducted between March 2019 and February 2020, with patients diagnosed with SCD. Patients were questioned about the perception of prejudice in any kind of situation, choosing between "Yes" or "No", not differentiating situations related to prejudice. To assess the QoL and impact of the disease, the volunteers answered a version of the SF-36 questionnaire translated and validated into Brazilian Portuguese.
Results: In this study, 113 patients with SCD were followed up, 92% were classified as HbSS and the rest, divided between HbSC and HbS-β-0. Regarding the SF-36, the worst scores were in the summary of the physical components (mean 48.19 ± 21.51) and the physical aspect had the lowest mean (30.75 ±€42.65). When questioned if they had already perceived any kind of prejudice, including the SCD, 32.74% answered "Yes". For this comparison, there was a significant difference in the summary of the physical and mental components, with worse QoL for those who had already suffered prejudice.
Conclusion: Patients diagnosed with SCD who reported perception of prejudice had statistically significant worse QoL, revealing the negative impact, that might lead to sadness and social isolation.

Introduction

The occurrence of labeling, stereotyping and prejudice in interpersonal relations and discrimination in the context of health situations is called Health-Related Stigma. This

* *Corresponding author at:* Universidade Federal Universal do Maranhão (CCBS UFMA), São Luís, MA, Brazil.
E-mail address: camila.freitas1101@gmail.com (C.F.d.A. Rodrigues).
https://doi.org/10.1016/j.htct.2021.06.002

scenario leads the patient to perceive a different treatment, due to the disease stigma.[1,2]

Patients with chronic diseases can suffer prejudice in different manners, leading to stigmatization. After all, they have different characteristics from those accepted in society and might be treated differently by the community, which may have misinterpretations and inaccurate beliefs about the stigmatized person.[3]

Sickle cell disease (SCD) is a hereditary hemoglobinopathy that affects millions of individuals worldwide, being one of the most frequent genetic alterations in Brazil.[4,5] The SCD is associated with complications, such as chronic anemia, acute and chronic pain, stroke, acute chest pain syndrome and long-term damage to target organs.[4] Although SCD is a genetic disease, being more prevalent in the brown and black population, it is impacted by racism and by issues of equity in health care, including difficult access to care and less financial support.[6]

It is a debilitating disease that results in a progressive decrease in health-related quality of life (QoL) over time due to the impact of disease-related complications,[1–6] generating a risk of stigmatization of the disease, which refers to a form of devaluation, judgment or social disqualification of individuals based on a health-related condition.[7,8] It can also lead to an inability to maintain a consistent professional or school activity, to participate in daily, social or recreational activities and to participate in family life.[9]

Pain episodes involving vaso-occlusion mechanisms are the most common complications in SCD, being the reason for most of the health system demand.[10] Pain episodes often increase with age and pain chronification occurs in 30–40% of adolescents and adults with SCD, significantly impairing the QoL.[8]

The perception of prejudice and stigmatization of the disease in SCD may be related to a worse quality of life because it is a chronic condition that imposes several limitations in the lives of carriers, associated with clinical characteristics and difficulty of access to treatment.[11,12]

This study aimed to describe and evaluate the perception of the prejudice against the disease and its impact on the quality of life of patients with sickle cell disease.

Material and method

Study design

This is a cross-sectional study conducted between March 2019 and February 2020 with sickle cell disease patients treated at the Hematology and Hemotherapy Center of Maranhão (HEMOMAR) in the city of São Luís, being the only reference for the treatment of hemoglobinopathies in the whole State of Maranhão, located in the northeast region of Brazil, characterized by being one of the poorest states and having the second lowest Human Development Index (HDI), Maranhão's being 0.639, while that of Brazil is 0.761.[13,14] This study was approved by the Research Ethics Committee of the Federal University of Maranhão.

Sample

The patients diagnosed with SCD were evaluated and followed up regularly at the specialized outpatient clinic of HEMOMAR. After an explanation about the study, the interviewee signed the Free and Informed Consent Form (ICF). Participants under the age of 18 also signed the ICF, as did their guardians.

Patients with SCD aged 14 or older, diagnosed by hemoglobin electrophoresis (high-efficiency liquid chromatography technique), with full physical, mental and intellectual conditions to communicate with the researchers and who agreed to participate in the investigation, were included. Patients under 14 years of age, patients with psychiatric disorders, hearing or speech disabilities and pain episodes at the time of the interview were excluded.

As of May 2018, there were approximately 700 patients registered at HEMOMAR with the diagnosis of SCD, of which approximately 250 were 14 years of age or over and could therefore be invited to participate in this study. Thus, authors have considered for the sample calculation a minimum of 15% difference in the QoL score between the users of hydroxyurea (HU), the standard treatment offered in Brazil, and non-users of this drug, yielding a sample of 111 patients, with a sampling error of 5% and a 95% confidence level.

Data collection instruments

Sociodemographic and clinical characteristics form

The volunteers were interviewed by the researcher to fill out a sociodemographic form and provide anthropometric data (weight and height).

Information was then collected to characterize the disease, such as age at diagnosis, classification of the disease, chronic complications, comorbidities, occurrence of pain episodes, perception of prejudice related to the disease, use of opioids, number of transfusions in the last 12 months and information on the use of hydroxyurea (HU). The patients were questioned about the perception of prejudice in any kind of situation, choosing answers between "Yes" or "No", not differentiating situations of prejudice related to ethnicity or health condition.

Laboratory data requested in the routine HEMOMAR consultation (hemoglobin concentration, leukocytes and platelet count, reticulocyte count, lactate dehydrogenase dosage and fetal hemoglobin level) were recorded when filling out the form.

Interview

Participants were approached at the end of their medical consultation with the hematologist for a recorded interview. The interview was conducted with open-ended questions addressing their experiences of perceived prejudice, perception of the disease and how it affected their daily lives since their childhood.

Quality of life questionnaire - SF-36

For the evaluation of QoL, the volunteers answered a version of the SF-36 questionnaire translated and validated into Brazilian Portuguese[15] to assess the impact of the disease on patients with SCD. The SF-36 is a multidimensional questionnaire with 36 items divided into 8 scales or domains: functional capacity, physical aspects, pain, general health, vitality, social and emotional aspects and mental health. These domains are divided into two components: physical and mental. Each item has a value between 0 and 100 points and the highest scores correspond to a better QoL. The scores are calculated according to the points of the items of each domain, as well as those of the components that are derivations from the related domains. The scores between 0 and 49 were classified as a bad quality of life, whereas those between 50 and 100 were considered a good quality of life.[15]

Statistical analysis

The data were analyzed by the GraphPad Prism 7® Program. The Kolmogorov-Smirnov test evaluated the normality of the data. The samples were compared using the Student's *t*-test for two groups; for more than two groups, the univariate analysis of variance (One-way ANOVA) was performed. The level of statistical significance used was 5% ($p < 0.05$).

Result

In this study, 113 patients with sickle cell disease were followed up at HEMOMAR were evaluated according to the inclusion criteria. General sociodemographic characteristics are described in Table 1.

Table 1 – Sociodemographic characteristics of sickle cell patients in São Luís, Maranhão, 2019.

Characteristics	N	Percentage (%)
Genre		
Male	42	37.2
Female	71	62.8
Age		
14 to 30 years	76	67.2
> 30 years	37	32.8
Marital status		
Not married	86	76.1
Married / stable relationship	26	23.1
Divorced	1	0.8
Race/Color		
White	9	7.9
Brown	64	56.7
Yellow	4	3.5
Black	36	31.9
Residence		
São Luís	22	19.5
Other	91	80.5
Education		
Incomplete elementary school	34	30
Complete elementary school	12	10.7
Incomplete high school	27	23.9
Complete high school	33	29.2
College Graduate	7	6.2
Religion		
Catholic	62	54.8
Evangelical	41	36.2
Without religion	10	9
Monthly family income		
<1 minimum wage	21	18.5
1 to 2 minimum wages	78	69.1
3 to 5 minimum wages	11	9.8
> 5 minimum wages	3	2.6

The most cited occupation among those surveyed was student (39.82%), followed by 30.9% of unemployed individuals, with the remaining varying among farmers, teachers and masons. Concerning the anthropometric measurements made during the interview, the average height among the patients was 159.9 cm., with an average weight of 50.72 kg.

As for the characteristics of the disease, 92% of the interviewees had SCD classified as HbSS, the rest being divided between HbSC and HbS-β-0. The diagnosis of SCD by newborn screening was made in 27.43% of the SCD patients; 19.46% of the patients had diagnostic confirmation by five years of life; 27.43% between 6 and 15 years, and; 25.55% only over 16 years of age.

Of the 113 individuals who answered the questionnaire, 48 (42.47%) have been using HU for more than 6 months. The average dose used was 24.16 mg/kg/day.

The complication most reported by the patients studied was chronic pain, present in 72.91% of HU users and 87.69% of those without HU. Secondly, priapism was observed in the male population, with rates of 63.15% in those who received HU and 56.52% in those without the drug. It is followed by acute chest syndrome, present in 45.83% of patients with the drug and 30.76% in those without. Cholelithiasis was also present with higher rates for those without HU (53.84%) against 41.66% in those with HU. Among the comorbidities questioned for those interviewed, 5.3% claimed to have systemic arterial hypertension; 4.4% had hepatitis B, and; 4% had an iron overload secondary to multiple transfusions.

The SF-36 quality of life questionnaire forms were first evaluated, calculating all the components of each domain and grouping them into physical components (functional capacity, physical aspects, pain and general health status) and mental components (vitality, social aspects, emotional aspects and mental health).

The means for each sub-domain of the SF-36 are represented in Table 2. It can be observed that the worst scores were represented by the summary of the physical components (mean 48.19 ± 21.51), highlighting among them the physical aspect as the one with the lowest means.

Separating patients into groups according to the use of hydroxyurea for the treatment of SCD, it was observed that the SF-36 scores were better in patients who used the medication, having statistical significance in the physical components ($p = 0.0101$) (Figure 1).

When questioned if they had already perceived any kind of prejudice, including the disease itself, 32.74% answered "Yes". For this comparison, there was a significant difference, both in the summary of the physical and mental components, with a worse quality of life for those who had already suffered this type of injury (Table 3).

The pain episodes were assessed over the 12-month period. For this, groups of patients were formed according to

Table 2 – General quality of life of patients with SCD, using the SF-36 questionnaire, in São Luís, Maranhão, 2019.

DOMAINS	Mean ± SD *n* = 113	Minimum value	Maximum value
Functional capacity	61.24 ± 22.93	10	100
Physical aspect	30.75 ± 42.65	0	100
Pain	64.98 ± 32.33	0	100
General health status	35.80 ± 20.47	0	90
Summary - Physical components	48.19 ± 21.51	10	98.75
Vitality	55.04 ± 23.17	0	100
Social aspects	85.18 ± 22.07	0	100
Emotional aspects	60.18 ± 45.83	0	100
Mental health	72.07 ± 22.58	8	100
Summary - Mental components	68.00 ± 22.32	6.5	100

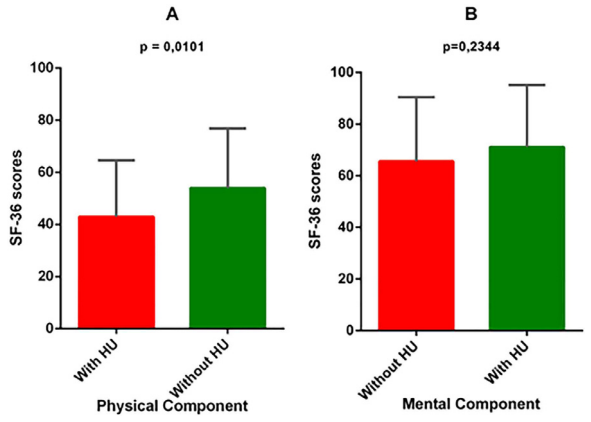

Figure 1 – SF-36 questionnaire scores and their distribution between the physical(A) and mental (B) componentes in patients who used hydroxyurea or not.Results expessed in mean ± SD.Values of p = 0.0101 between physical domains and p = 0.2344 between mental domains, considering patients who used hydroxyurea or not. Student's test.

the frequency of pain in this period. No episodes were reported in 12 patients; one to two episodes in 41 individuals, and; three or more episodes in 60 SCD patients. The impact of pain can be demonstrated in the inversely proportional relationship between the QoL and the number of pain episodes, with a statistically significant *p*-value, both for the summary of the physical components and for the mental components (Table 4).

To evaluate the impact of blood transfusions on the QoL, patients were divided into three groups, considering the last 12 months: the first with patients who did not receive a transfusion; the second with those who received less than ten units of RBC concentrate, and; the third with those who received ten or more units. It was observed that QoL was better in those who did not receive blood units, but without statistical significance. An important aspect to be highlighted is that the sub-domain pain had a *p*-value of 0.002 (Table 5)

The number of hospitalizations in the last 12 months had no impact on the summary of physical and mental components. However, it is worth noting that the sub-domain pain had a statistically significant difference, demonstrating that the occurrence of such symptoms affects the perception of comfort and the QoL of the patients (Table 6).

Among the recorded and transcribed interviews, the most relevant and recurrent statements were used to contextualize the problem. Furthermore, some patients were not comfortable talking about the details of the prejudice issues.

Patient A: *"I was never picked on the soccer team because I got tired so easily! I didn't like that"*.

Patient B: *"The teacher always sent me back home saying I had hepatitis!"*.

Table 3 – Quality of life and prejudice against SCD evaluated by the SF-36 questionnaire in São Luís, Maranhão, 2019.

QUALITY OF LIFE DOMAINS (mean ± SD)	PREJUDICE		*p**
	No *n* = 76	Yes *n* = 37	
Functional capacity	64.67 ± 22.28	54.19 ± 22.93	0.0219
Physical aspect	41.45 ± 45.38	8784 ± 25.15	<0.001
Pain	75.86 ± 27.76	42.97 ± 29.56	<0.001
General health status	37.83 ± 19.77	31.62 ± 21.51	0.1310
Summary - Physical components	54.52 ± 22.60	33.59 ± 15.45	<0.001
Vitality	58.95 ± 23.07	47.84 ± 20.23	0.0140
Social aspects	88.82 ± 20.77	77.70 ± 23.04	0.0114
Emotional aspects	63.16 ± 44.75	54.05 ± 48.02	0.3239
Mental health	75.53 ± 21.68	64.97 ± 23.01	0.0190
Summary - Mental components	72.07 ± 23.75	59.40 ± 24.28	0.0095

* Student's *t*-test.

Table 4 – Quality of life and occurrence of pain episodes in patients with SCD evaluated by SF-36 questionnaire in São Luís, Maranhão, 2019.

	PAIN EPISODES			
QUALITY OF LIFE DOMAINS (mean ± SD)	None $n = 12$	1 or 2 episodes $n = 41$	≥ 3 episodes $n = 60$	p^*
Physical function	80.42 ± 27.75	61.95 ± 20.40	56.92 ± 21.86	0,.043
Physical role	56.25 ± 46.62	39,02 ± 47.45	20.00 ± 34.99	0.0071
Pain	87.71 ± 23.92	75.06 ± 28.84	53.08 ± 32.14	< 0.001
General health perceptions	45.00 ± 26.02	39.39 ± 16.51	31.50 ± 20.96	0.0407
Summary – Physical components	68.47 ± 21.03	53.61 ± 22.03	39.44 ± 19.67	0.0001
Vitality	75.00 ± 15.95	58.17 ± 20.12	49.42 ± 23.16	0.0008
Social role functioning	89.58 ± 12.87	89.33 ± 17.36	81.46 ± 25.08	0.1630
Emotional role functioning	83.33 ± 33.34	58.53 ± 47.01	56.67 ± 46.46	0.1773
Mental health	86.33 ± 8.60	76.49 ± 18.81	66.20 ± 24.98	0.0047
Summary – Mental components	84.69 ± 13.82	71.51 ± 19.43	62.12 ± 27.44	0.0064

*One-way ANOVA.

Table 5 – Quality of life and transfusions received in 12 months by patients with SCD by SF-36 questionnaire in São Luís, Maranhão, 2019.

	TRANSFUSIONS			
QUALITY OF LIFE DOMAINS (mean ± SD)	None $n = 39$	< 10 units $n = 43$	≥ 10 units $n = 31$	p^*
Physical function	67.69 ± 20.74	60.93 ± 19.53	53.55 ± 27.72	0.0357
Physical role	35.90 ± 44.72	32.56 ± 43.48	21.77 ± 38.59	0.3679
Pain	75.83 ± 26.96	61.74 ± 35.90	56.53 ± 30.06	0.0288
General health perceptions	38.08 ± 20.79	32.67 ± 19.50	37.26 ± 21.48	0.4437
Summary - Physical components	53.80 ± 22.38	46.69 ± 22.41	41.30 ± 22.29	0.0675
Vitality	58.08 ± 22.90	51.86 ± 21.35	56.61 ± 24.34	0.4367
Social role functioning	93.59 ± 14.01	83.72 ± 21.57	76.61 ± 27.34	0.0044
Emotional role functioning	70.08 ± 44.46	55.04 ± 45.36	54.84 ± 47.57	0.2505
Mental health	76.62 ± 19.37	71.07 ± 20.45	67.74 ± 28.19	0.2479
Summary - Mental components	76.62 ± 19.37	65.72 ± 23.42	63.13 ± 27.56	0.1318

* One-way ANOVA.

Table 6 – Quality of life and hospitalizations of patients with SCD by SF-36 questionnaire in São Luís, Maranhão, 2019.

	HOSPITAL ADMISSIONS		
QUALITY OF LIFE DOMAINS (mean ± SD)	< 3 $n = 93$	≥ 3 $n = 20$	p^*
Physical function	61.88 ± 23.08	58.25 ± 22.55	0.5229
Physical role	31.72 ± 42.85	26.25 ± 42.52	0.6050
Bodily pain	69.30 ± 30.89	46.48 ± 31.95	0.0035
General health perceptions	36.88 ± 20.89	30.75 ± 18.01	0.2258
Summary - Physical components	49.51 ± 22.18	39.08 ± 23.80	0.0623
Vitality	54.95 ± 22.95	57.00 ± 21.97	0.7153
Social role functioning	86.56 ± 18.91	78.75 ± 32.97	0.1520
Emotional role functioning	60.93 ± 45.48	56.67 ± 48.49	0.7078
Mental health	72.77 ± 21.89	68.80 ± 25.91	0.4776
Summary - Mental components	68.45 ± 23.90	65.45 ± 27.94	0.6218

* Student's *t*-test.

Patient C: *"I don't know if, for me, the worst thing was the prejudice with the color of my skin or the sickle cell anemia"*.

Patient D: *"The most annoying thing is being left out of what I enjoy doing the most"*.

Discussion

Chronic diseases can lead to the development of physical, social and emotional problems due to organic dysfunctions of

the underlying disease. In this scenario, SCD also presents such characteristics. These complications can occur in its natural course, interfering directly in the quality of life, and can be related to the perception of stigmatization and prejudice of patients diagnosed with SCD.

Although SCD was initially described over a century ago, the development of disease-modifying therapies has stagnated due to inadequate research funding, at least in part attributable to structural racism.[16,17] In addition to the substantial barriers created by prejudice, the access to and delivery of high-quality health care for patients with SCD is also disrupted by interpersonal racism. Too often patients with SCD simultaneously must deal with unbearable pain and racist attitudes expressed by healthcare workers.[16,18]

The SCD is more prevalent in the population of African descent in Brazil, which corresponds to more than 50% of the Brazilian population, according to data from the Brazilian Institute of Geography and Statistics, but on the other hand, it is part of the most socially and economically vulnerable segments of the country.[5,19,20]

As the pain crises begin in childhood and, as verified in this study, constitute the most prevalent complications in this population, these youngsters learn to deal with the disease and the limitations imposed by it from an early age. When dealing with young people with SCD, hospitalizations limit their satisfactory school development, also making them stigmatized as sick and absent students. Moreover, SCD can create limitations in the practice of sports. This fact is fundamental because sports are important instruments of social insertion, mainly for the population with a higher social risk. Due to all these factors, it is necessary to maintain a multidisciplinary team which provides psychological and social support throughout the school life of this population.[17]

Another aspect that represents a negative impact, leading to sadness and social isolation, is the prejudice and discrimination suffered by such patients. As demonstrated in this research and in another study,[11] SCD carriers had the perception that their QoL was impaired, both in physical and mental aspects. This reality constitutes a deeply worrisome picture for most people with SCD, since it deteriorates their self-esteem, leading to the construction and perception of a negative image of themselves.[7,18]

The term “sickler” and its culture-specific variations, a term connected with racist origins, is often used ignorantly to describe and depersonalize patients with SCD.[16,21] Also, even in the era of opioid pandemics, patients with SCD are often described as drug seekers and accused of feigning their pain, which results in inadequate treatment and more suffering.[21,22]

Because of the challenges in receiving adequate care and the stress associated with perceived racial stigma, many patients choose to avoid care altogether, further increasing the risk of life-threatening complications.[17] Despite suffering from severe pain, patients report getting dressed nicely before presenting themselves to the emergency department in an attempt to avoid judgment and receive better care.[7,23]

In this study, the most frequent complaints were related, for example, to the exclusion from sports activities by teachers and other partners, because they thought the patients had little resistance, or also they were prevented from attending school due to jaundice, misinterpreted as hepatitis or an infectious disease.

Although there are many sources of stigma experienced by persons with SCD, racism is an important source, as most affected individuals are black or brown. Racism frequently interacts and exacerbates other sources of health-related stigma in SCD, including disease- and opioid-based stigmas. This context leads to racism being recognized as a source of stigma or akin to it. Despite SCD being a genetic disease, it is impacted by racism and health care equity issues, including hindered access to care and less funding support.[24,25]

A systematic review demonstrated the impact that stigma has on the life and health of individuals with SCD, hindering physical and psychological well-being, having negative social consequences and damaging interactions with health services.[7] Despite being a prevailing condition, the general population is often not educated about SCD and ultimately forms its own opinions, usually negative, about people with SCD. This results in people with SCD being devalued and suffering a loss of status. They also have their experiences of pain discredited by others and are accused of being weak, lazy, or pretending to be sick.[11,26]

Although few studies have evaluated the QoL of people with SCD, they have disclosed that such patients have a worse health-related QoL than the population without the disease.[27] Interest in measuring the QoL has become fundamental in healthcare practices, from disease prevention to health promotion. Thus, the evaluation of the QoL has become an important area of scientific knowledge because it allows for a more objective and clear evaluation of the global impact of chronic diseases, such as SCD.

The evaluation of the QoL of people with SCD can reflect the advances of science in the care of SCD, available in the Unified Health System (SUS), a universal and free Brazilian public health system that provides attention and care to the entire population. In addition, the country has a National Policy of Integral Care for People with Sickle Cell Disease, available to all these people, which contributes to the greater goal that is longevity with quality of life and can aggregate and improve the results of research in this area.[19,20]

The average of the summaries of the physical and mental components of the SF-36 questionnaire among all interviewees (48.19 and 68.00, respectively) revealed that the physical commitment is more pronounced than the mental one. A similar study conducted in Cameroon in 2017 showed averages of 47.3 and 41.0, respectively, for the physical and mental components, suggesting worse values in the latter, different from those presented in this study, which could be explained by the conditions of sociopolitical instability there.[28] Another study in Saudi Arabia revealed even lower values than those in the Cameroonian study, probably because the SF-36 was also applied to patients during pain episodes.[29,30] Moreover, studies with different methods of evaluating the QoL, have shown an important impact of SCD on the QoL, in addition to the association of factors, such as frequency and intensity of pain episodes, with lower QoL values.[29,30]

The domain of the physical aspect, one of the items of the SF-36 physical component, is composed of four questions (related to problems at work or some daily activity due to his/her physical health). Regarding issue four of this domain,

which refers to problems with some regular daily task as a consequence of health, it was observed that a large majority of the population decreased the amount of time they dedicated to work, performing fewer tasks than they would like, and limiting their type of work or their tasks, as well as mentioning difficulties in performing their work or other occupations. Thus, this group of patients with SCD presented a momentary inability to develop their activities, working or not, a fact that deserves attention, since they are at a productive age for society.

The contribution of this research in the evaluation of the advances of science in the care of SCD available in the public health system is necessary and can be added to the actions of the National Policy of Integral Care for People with SCD. Integrality, one of the SUS principles, states that "The user is the center of the line of care" and therefore the attention and care, especially for those with chronic diseases, must always be evaluated by looking at the quality of life that science can produce and the identification of vulnerable points of care that do not lead to this end.[19,20]

A limitation of this cross-sectional study is that chronic complications have been reported by patients, depending exclusively on the history of the interviewee, and may generate bias due to forgetfulness, or even lack of knowledge about their complications. It should also be emphasized that only one treatment center was studied, with its own economic and social context, which may be different from other centers, including those within the country itself. As for the query concerning the perception of prejudice in general, the researchers considered that the different situations involving prejudice perception add up to the manner and intensity in which it affects the patients.

Conclusion

To date, there are few specific instruments to assess the QoL of people with SCD, but some generic instruments serve this purpose. New research is needed to increase understanding of the impact of prejudice perception on the quality of life of adults with SCD and may be useful in promoting health and indirectly influencing the state of the disease. Because their QoL is impaired by the disease, patients with SCD need public policies adopted and continuously reviewed to manage their condition.

Conflicts of interest

The authors declare no conflicts of interest.

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