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ORIGINAL ARTICLE

QUALITY OF LIFE OF SICKLE CELL DISEASE CARRIERS QUALIDADE DE VIDA DOS PORTADORES DE DOENÇA FALCIFORME CALIDAD DE VIDA DE LOS PORTADORES DE ENFERMEDAD FALCIFORME

Kamila Tuany Lacerda Leão Lima¹, João Otávio Ferreira Pereira², Paulo Roberto de Melo Reis³, Keila Correia de Alcântara⁴, Flávia Melo Rodrigues⁵

ABSTRACT

Objective: to evaluate the quality of life of sickle cell patients treated by the Sickle Cell Anemia Program. **Method:** this is a quantitative, descriptive and analytical study in 20 patients with sickle cell anemia and 40 non-sickle individuals. Data was collected through interviews between February and May 2015. Quality of life was evaluated through SF-36 and WHOQOL-BREF. Results were presented in the form of tables. **Results:** the majority of patients with sickle cell disease declare themselves as black and brown with a low level of schooling; the physical aspects and the functional capacity had the worst results and, with age, the physical aspect becomes more compromised. The SF-36 questionnaire showed that, among the scores, individuals with FD presented pain, functional capacity, vitality, physical, emotional and mental health aspects as the most impaired in relation to the group of patients without SCD. **Conclusion:** the WHOQOL-BREF evaluation showed a significant impairment of physical and general quality of life among patients with DF; participants with sickle-cell disease have a negative impact on quality of life, which interferes with and influences the health of these people. **Descritores:** Quality of Life; Chronic Disease; Sickness Impact Profile; Sickle Cell Anemia; Anemia Hemolytic; Erythrocytes.

RESUMO

Objetivo: avaliar a qualidade de vida de portadores de células falciformes atendidos pelo Programa de Anemia Falciforme. **Método:** trata-se de um estudo quantitativo, descritivo e analítico em 20 portadores de anemia falciforme e 40 indivíduos não falciformes. Coletaram-se os dados por meio de entrevistas no período entre fevereiro a maio de 2015. Avaliou-se a qualidade de vida por meio de SF-36 e WHOQOL-BREF. Apresentaram-se os resultados em forma de tabelas. **Resultados:** constata-se que a maioria dos pacientes com doença falciforme se declara como negros e castanhos e com baixo nível de escolaridade; os aspectos físicos e a capacidade funcional tiveram os piores resultados e, com a idade, o aspecto físico se torna mais comprometido. Mostrou-se, pelo questionário SF-36, que, entre os escores, os indivíduos com DF apresentavam dor, capacidade funcional, vitalidade, aspectos físicos, emocionais e de saúde mental como os mais prejudicados em relação ao grupo de pacientes sem DF. **Conclusão:** apresentou-se, pela avaliação WHOQOL-BREF, comprometimento significativo da qualidade de vida física e geral entre os pacientes com DF; já os participantes com doença falciforme sofrem um impacto negativo na qualidade de vida, o que interfere e influencia a saúde dessas pessoas. **Descritores:** Qualidade de Vida; Doença Crônica; Perfil de Impacto da Doença; Anemia Falciforme; Anemia Hemolítica; Eritrócitos.

RESUMEN

Objetivo: evaluar la calidad de vida de portadores de células falciformes atendidos por el Programa de Anemia Falciforme. **Método:** se trata de un estudio cuantitativo, descriptivo y analítico en 20 portadores de anemia falciforme y 40 individuos no falciformes. Se recogieron los datos a través de entrevistas en el período entre febrero a mayo de 2015. Se evaluó la calidad de vida por medio de SF-36 y WHOQOL-BREF. Se presentaron los resultados en forma de tablas. **Resultados:** se constata que la mayoría de los pacientes con enfermedad falciforme se declara como negros y castaños y con bajo nivel de escolaridad; los aspectos físicos y la capacidad funcional tuvieron los peores resultados y, con la edad, el aspecto físico se vuelve más comprometido. Se mostró, por el cuestionario SF-36, que entre los escores, los individuos con DF presentaban dolor, capacidad funcional, vitalidad, aspectos físicos, emocionales y de salud mental como los más perjudicados en relación al grupo de pacientes sin DF. **Conclusión:** se presentó, por la evaluación WHOQOL-BREF, un compromiso significativo de la calidad de vida física y general entre los pacientes con DF; ya los participantes con enfermedad falciforme sufren un impacto negativo en la calidad de vida, lo que interfiere e influye en la salud de esas personas. **Descritores:** Calidad de Vida; Enfermedad Crónica; Perfil de Impacto de la Enfermedad; Anemia de Células falciformes; Anemia Hemolítica; Eritrocitos.

¹Master, University Center FG / CESC, Guanambi (BA), Brazil. Email: kamilatl@gmail.com ORCID iD: <http://orcid.org/0000-0001-6547-3501>; ²Biomedic, Guanambi (BA), Brazil. E-mail: joaotavio_gbi@hotmail.com ORCID iD: <http://orcid.org/0000-0003-0696-7848>; ³PhD, Pontifical Catholic University of Goiás / PUCGO, Goiânia (GO), Brazil. Email: melo_reis@yahoo.com.br ORCID iD: <http://orcid.org/0000-0001-9660-2572>; ⁴PhD, Federal University of Goiás / UFG, Goiânia (GO), Brazil. Email: keilalcantara@yahoo.com.br ORCID iD: <https://orcid.org/0000-0002-4477-2833>; ⁵PhD, Pontifical Catholic University of Goiás / PUCGO, State University of Goiás / UEG, Goiânia (GO), Brazil. Email: rflamelo@gmail.com ORCID iD: <https://orcid.org/0000-0002-2557-6570>.

INTRODUCTION

Sickle cell disease (SCD) is defined as a set of different genotypes characterized by the presence of hemoglobin S (HBS), a variant of normal hemoglobin, characterized by the substitution of the sixth amino acid beta globin; glutamic acid by valine. This HBS polymerization is carried out under low oxygen pressures, altering the shape of the red blood cells and shortening their average life time, thus causing problems such as vaso-occlusion crisis and, consequently, organ damage.¹⁻³

SCD can be presented in different ways: heterozygosis, in which the hemoglobin S gene may interact with other variant hemoglobins or, also, associate with normal adult hemoglobin, hemoglobin A, characterizing the asymptomatic carrier with known benign condition as a sickle cell trait. There is the homozygous form, represented by hemoglobin S, known as sickle cell anemia, which is the most clinically severe form of the disease.⁴⁻⁵

It is known that sickle cell disease was brought to the Americas by the immigration of African peoples, and its distribution in Brazil occurs in a heterogeneous way, where the most affected region is characterized by the largest Afrodescendant population, which, for the most part, inhabits poorer and poorer places, where their own environmental aspects influence the pathophysiology of the disease. It is reported that the most affected regions are North and Northeast. A prevalence of one case of sickle cell disease is reported for Bahia, the State chosen for this study, for every 650 live births, while in the State of Rio Grande do Sul the prevalence is one case for every 11 thousand live births.^{2,6-7}

It is noted that SCD is a chronic disease, which brings some limitations in daily life and changes in the lifestyle changes of affected individuals.⁸

SCD patients can now benefit from the cure when undergoing bone marrow transplantation. This treatment is used because it is feasible and prolong the life of children with sickle cell anemia, however, it is an aggressive procedure, which can compromise multiple organs and cause immunological depression; therefore, questions about the quality of life arise in the face of this situation because, on the one hand, the treatment guarantees the prolongation of life, on the other, it is difficult to be faced by the patient and his family due to the complexity of this process.⁹

The chronic disease directly affects the quality of life of its patients, which is the life

condition of an individual when assessing the impacts of the disease on the commitment of daily activities, emotional, functional, psychosocial and life expectancy.¹⁰ It is something that is individual, only possible to be evaluated by the subject itself.¹¹

OBJECTIVE

- To evaluate the quality of life of sickle cell patients treated by the Sickle Cell Anemia Program.

METHOD

This is a quantitative, descriptive and analytical study carried out in the Guanambi micro-region, Bahia, including five municipalities of the eighteen that compose the studied area: Guanambi, Iuiú, Riacho de Santana, Palmas de Monte Alto and Pindaí. The study was carried out with two groups, the case and the control, and the case group was represented by patients with sickle cell disease attended at SCAP, between February and May 2015, aged 12 years or older, independently of sex. For the control group, people who did not present any chronic disease that could interfere directly with quality of life, but with sociodemographic and economic characteristics similar to those of the participants in the case group, such as age, family income and municipality source were included.

The SF-36 and the WHOQOL-BREF, which are tools for the assessment of quality of life, and a sociodemographic and economic questionnaire prepared by the researchers, were used for data collection. Fisher's exact test was used to compare socioeconomic and demographic variables in both groups. Before choosing which mean comparison test to apply, the data using the Kolmogorov-Smirnov test were analyzed to see if they followed normal distribution. The Student t and Mann-Whitney t tests were used to compare the income and age of participants with sickle cell disease and without sickle cell disease. In order to verify if there was a significant difference between the means of the scores of the two instruments (SF-36 and WHOQOL-BREF), in the two groups, Student's t test was performed. Spearman's correlation was used to evaluate the association between the scores of the two instruments, in addition to correlating with age.

It was obeyed, by the study, to the ethical precepts according to resolution 466/12, with the approval in the REC under the opinion of number 889.040.

RESULTS

A total of 20 patients with sickle cell disease were evaluated, being these heterozygotes associated with other hemoglobinopathies (C, thalassemia beta) and homozygotes (SS), and 40 participants without sickle cell disease in this study.

It was revealed that the majority were female in both groups, and those with SCD had lower schooling than the healthy group ($p = 0.0202$), with black skin color being more frequent in the SCD group than in the group without SCD ($p = 0.0135$) (Table 1).

Table 1. Sociodemographic characteristics of participants with SCD and without SCD. Guanambi (BA), Brazil, 2015.

Variables	With SCD n (%)	Without SCD n (%)	p^*
Gender			
Male	9 (45)	9 (22)	0.1335
Female	11 (55)	31 (78)	
Education			
With schooling	17 (85)	40 (100)	0.0202
Without schooling	3 (15)	0	
Homeowner			
Yes	17 (85)	30 (75)	0.5128
No	3 (15)	10 (25)	
Skin color			
Black	8 (40)	4 (10)	0.0135
Others	12 (60)	36 (90)	

*Fisher's Exact Test SCD: sickle cell disease

In the SF-36 questionnaire, eight domains investigate different aspects such as functional capacity, physical aspects, pain, general health, vitality, social aspects and mental health, and each domain received a value between zero and 100, which corresponds from the worst to the best state of health.

It was revealed that the general health status and the social aspect were not different between those with SCD and those

without SCD, however, patients with SCD had lower quality of life than those without SCD, when the functional capacity dimensions ($p < 0.0001$); physical aspects ($p = 0.0001$); pain ($p = 0.0365$); vitality ($p = 0.0045$); ($p = 0.0223$) (Table 2), and the emotional aspect and functional capacity were the dimensions with the worst scores in patients with SCD.

Table 2. Comparison of mean scores of the SF-36 dimensions between patients with SCD (n = 20) and without SCD (n = 40). Guanambi (BA), Brazil, 2015.

Dimensions SF-36	With SCD (n=20)	Without SCD (n=40)	p^*
	Average ($\pm$ sd)	Average ($\pm$ sd)	
Functional capacity	40.50 ($\pm$ 17.98)	80.12 ($\pm$ 19.67)	< 0.0001
Physical aspects	46.25 ($\pm$ 39.96)	82.50 ($\pm$ 27.85)	0.0001
Pain	51.30 ($\pm$ 27.58)	67.20 ($\pm$ 26.91)	0.0365
General health status	53.35 ($\pm$ 19.59)	58.23 ($\pm$ 17.05)	0.3247
Vitality	51.75 ($\pm$ 21.72)	68.00 ($\pm$ 19.28)	0.0045
Social aspect	76.88 ($\pm$ 21.94)	75.31 ($\pm$ 24.92)	0.8128
Emocional aspect	38.32 ($\pm$ 40.86)	68.32 ($\pm$ 38.46)	0.0071
Mental Health	53.00 ($\pm$ 24.96)	68.00 ($\pm$ 22.52)	0.0223

*Student's t test, sd = Standard Deviation

It was found that the mean age of the studied groups was 30 years, and the median family income was R\$ 788.00; of the SF-36, the physical aspect correlated negatively with

age ($p = 0.0238$), showing that the more advanced the age, the worse the general physical state (Table 3).

Table 3. Correlation between SF-36 dimensions and age of patients with SCD. Guanambi (BA), Brazil, 2015.

Dimensions SF-36	AGE	
	R	p*
Functional capacity	-0.1879	0.4276
Physical aspects	-0.5029	0.0238
Pain	0.0688	0.7731
General health status	-0.3169	0.1733
Vitality	-0.3914	0.0878
Social aspect	-0.2055	0.3848
Emocional aspect	-0.3175	0.1725
Mental Health	-0.1171	0.6229

*Spearman's Correlation

It is explained that the WHOQOL-BREF is a questionnaire developed by the World Health Organization that evaluates indicators of Quality of Life through its physical, social, psychological and environmental aspects.

In order to determine the scores related to the domains of the instrument, all the items of the domain referenced, dividing them by the mean and then multiplying them by four. At the second moment of each domain, the value of four was decreased and multiplied by

6.25 or 100/16. In the final result, values ranging from zero to 100 were reached, and the closer to 100, the better the quality of life.

Only the physical and quality of life aspects obtained a lower score in the group with SCD, however, none of the WHOQOL-BREF domains correlated with the age of the participants in the group ($p < 0.05$) (Table 4) with SCD, which could be observed through the SF-36 questionnaire.

Table 4. Comparison of mean scores of the WHOQOL-BREF dimensions between patients with SCD and without SCD. Guanambi (BA), Brazil, 2015.

Domains of WHOQOL-BREF	With SCD (n=20)	Without SCD (n=40)	p*
	Average (±sd)	Average (±sd)	
Physical	52.32 (±14.39)	73.22 (±17.10)	< 0.0001
Psychological	61.68 (±15.92)	68.75 (±12.76)	0.0678
Social	64.17 (±12.71)	66.67 (±18.97)	0.5464
Environment	53.45 (±11.99)	57.43 (±11.77)	0.2244
Total Quality of Life	57.91 (±11.45)	66.51 (±12.27)	0.0112

*Student's t test, sd = Standard Deviation

It is added that no significant associations between the domains of the two instruments were found (Table 5).

Table 5. Correlation between SF-36 dimensions and WHOQOL-BREF domains among participants with sickle cell disease (SCD). Guanambi (BA), Brazil, 2015.

Dimensions SF-36	DOMAINS OF WHOQOL-BREF		
	PHYSICAL	PSYCHOLOGICAL	SOCIAL
Physical aspects	0.5586*	---	---
Mental health	---	0.6773*	---
Social aspects	---	---	0.5484*

*Spearman's Correlation (p)

DISCUSSION

It has been demonstrated by the results that the quality of life of patients with SCD is perceived by them as a poor quality, especially when related to physical and social aspects. The need to apply more than one

instrument for the assessment of quality of life was also evidenced in this study, since no correlation was found between the two.

The population group has been evaluated as historically less favored, because it is a region with a greater number of Afro-descendants and unfavorable socioeconomic

situation, which may reflect the low level of education and may also be associated with the limitations that the disease imposes.¹²

It is limited, due to the lack of knowledge about sickle cell disease, the participation of SCD carriers in social groups, as in the school environment, and this makes it difficult for them to form a personality and maintain their social relations.¹³

It is warned that, even in world terms, if one does not understand the environment in which one lives, the conditions of schooling, work and social life, in general, one has a small vision of the disease and, consequently, the solutions for improvement will have little effectiveness.¹⁴

It is understood that functional capacity is something that refers to the individual's ability to perform the basic physical and mental activities of the day to day, which showed a poor index among participants with sickle cell disease, and this may occur as a function of the constants vasoconstriction processes that may lead to complications of various organs, and such alterations reduce the functional capacity of patients with SCD, especially patients with sickle cell disease.¹⁵

The patient's physical condition is affected by SCD, possibly due to the symptoms characteristic of the disease, presenting dependence on treatment, susceptibility to fatigue and pain.¹⁶ The pain has a great negative impact on the quality of life of patients with SCD and even of their relatives, and their evaluation is very useful in clinical practice when demonstrating the extent of the problem.¹⁷ It was pointed out in a study that, due to pain, some domains related to the physical component of the SF-36 (functional capacity, pain, physical aspects and general health state) presented a significant reduction, and this shows that the domain limitation by aspects can be directly affected due to the pain felt by the patient.¹⁸

It was possible to evaluate the impact of psychological aspects on the well-being of the individual, the worst mean among the SF-36 domains, showing that this factor has great impacts on the patient's quality of life with SCD. It is inferred that the impairment of the domains social aspects and limitation by emotional aspect do not seem to be associated with the pain felt by the patients, but are influenced by other components of the disease. These impacts can be explained by the instability generated by the disease, linking them to the often unfavorable socioeconomic profile.¹⁸

In the field of mental health, questions about the state of mind, temperament, anxiety and psychological well-being are addressed, and the complications of the disease can generate psychological disorders, such as depression, presenting a compromise in the quality of life when associated with the symptomatology of this disorder.¹⁹

It is understood that with the advancement of the age, it is common to appear some factors that can interfere in the quality of life, such as instability, vulnerability to the limitations imposed by diseases, intellectual difficulty, including the gradual loss of its role in society until then performed. These factors negatively affect the quality of life, making the individual more dissatisfied with their condition.²⁰⁻¹

It is accepted that one of the most important points associated to the research objective was the choice of the instrument, since it had to be related to a cultural context compatible with that of the population studied,²² and, due to the specificities, this study used the two instruments to try to minimize their important limitations. The WHOQOL-BREF is based on proposing indicators that fail to assess the specificities of each subject in each context of assessment in the WHO quality of life (QoL) design, addressing aspects that comprehensively cover QoL, including factors that are or are not related to health, that is, evaluate the general quality of life. The SF-36 was developed to evaluate the aspects of health and activities generally affected by health conditions, only assessing the health-related quality of life, that is, more used for health-related quality of life assessment, more specifically.²³ Therefore, it is possible to observe very weak or insignificant correlations between the domains of these two questionnaires, as demonstrated in this study.

As an important limitation, the present study shows the size of the sample, which, when present in a reduced quantity, allows to consider the results only for the population in question.

CONCLUSION

It was possible to emphasize, by this study, that the participants with sickle-cell disease suffer a great negative impact on the quality of life, being, for the most part, blacks and with lower level of schooling.

It should be noted that the emotional and physical aspects and functional capacity were the dimensions of SF-36 and WHOQOL-BREF with worse scores in patients with SCD, and,

with advancing age, the physical aspect is compromised.

It was observed, in the correlation of the two instruments used in the study, that there was no correlation between them. It is necessary to direct the intended study, by the choice of instrument, to an evaluation of the quality of life related to health or in general, since the instruments studied here show differences when tested for the quality of life directed to health aspects.

It is pointed out by the research, the importance of the analysis of the quality of life, showing that it interferes and is also influenced by the health of patients with sickle cell disease.

The Sickle Cell Anemia Program (SCAP) was created in Guanambi (BA) in 2013 and currently serves 58 patients with sickle cell disease and also with other anemias that require follow-up. The SCAP's contribution to the quality of life of SCD patients is essential, who are regularly treated by qualified professionals.

It is concluded that the need exists for the expansion of the multiprofessional team of the Sickle Cell Anemia Program of the municipality of Guanambi, so that it can work more aspects that influence the quality of life, such as emotional ones, and that the health professionals are aware of these conditions, aiming to deal, in the best way, with the aspects that negatively interfere in the quality of life. This is more relevant in the case of chronic diseases, since it can directly influence the prognosis of the disease.

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Corresponding Address

Kamila Tuany Lacerda Leão Lima
Rua Presidente Costa e Silva, 100
Bairro Bela Vista
CEP: 46430000 – Guanambi (BA), Brazil