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Quality of Life of Sickle Cell Disease Patients of a Micro-Region in the Southwest of the State of Bahia (Brasil)

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Abstract

Sickle cell disease is understood as a set of different genotypes characterized by the presence of hemoglobin S and recognized as a chronic disease that can reduce the carrier's quality of life.

Objective: The study mainly aimed to evaluate the quality of life of sickle cell disease carriers of the micro-region of Guanambi, State of Bahia, attended by the Sickle Cell Anemia Program.

Method: A quantitative-descriptive-analytical study was conducted on 20 sickle cell carriers and 40 non-Sickle cell disease individuals. Data were collected through interviews to gather sociodemographic and economic information. Quality of life was assessed through SF-36 and WHOQOL-BREF.

Results: The sickle cell disease group had more dark skinned individuals (p=0.0033) and lower schooling than the non sickle cell disease group (p=0.0055). The SF-36 questionnaire showed that sickle cell disease carriers had more pain (p=0.0223), lower functional capacity (p=0.0001), vitality (p), physical (p), emotional and mental health aspects disadvantage compared with the non sickle cell disease group (reword). The assessment through WHOQOL-BREF showed significant impairment of the physical and total quality of life components among sickle cell disease carriers. There was no association between SF-36 and WHOQOL-BREF components.

Conclusion: Sickle cell disease participants suffer a negative impact on the quality of life, which interferes and is influenced by patients' health.

Keywords: Quality of life, Chronic disease, Sickle cell anemia, SF-36, WHOQOL-BREF

Introduction

Sickle cell disease is a set of different genotypes characterized by the presence of hemoglobin S, a variant of normal hemoglobin, featured by the replacement of the sixth beta-globing amino acid glutamic acid with valine [1,2]. This HbS undergoes polymerization under low oxygen pressures, altering the shape of red blood cells and shortening their average lifetime, thus ensuing problems such as vaso-occlusive crisis and, consequently, injuring organs [3] Sickle cell disease may manifest itself in different ways: heterozygosis, known as sickle cell trait and represented by the presence of hemoglobin A and S; associated with other hemoglobinopathies, such as C, D and Beta thalassemia; or in homozygous form, represented by hemoglobin S, known as sickle cell anemia, the most severe form of the disease clinically speaking [4-11].

It is known that sickle cell disease was brought to the Americas by the inflow of African people. It is heterogeneously spread in Brazil, where the most affected region is characterized by the largest Afro-descendant population, mostly living in poorer and needier locations where the very environmental aspects influence the path physiology of the disease. The most affected areas are the northern and northeastern regions give percentage compared to other regions. The State of Bahia chosen for this study has a prevalence of one sickle cell disease case per 650 live births [1,12,13].

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Sickle Cell Disease (SCD) is a chronic disease that ensues some constraints which to daily life and changes in the lifestyle of affected individuals. It also impacts social groups with varying levels of severity [14-17].

Treatment of chronic diseases includes changes in the lifestyle of patients and their continuous monitoring, with no claim to cure. It is one of the main reasons for seeking health services [17,18]. Sickle cell disease carriers can now benefit from healing when submitted to bone marrow transplant. Such treatment is used because it is feasible and extends the lives of children with sickle cell disease, but is an aggressive procedure which can affect multiple organs and cause immunosuppression. Thus, quality of life issues arise in this situation, since, on the one hand, this treatment ensures the continuation of life and, on the other hand, causes hardship for patients and their families due to the complexity of this process [19].

Chronic disease affects directly the quality of life of its patients, which is the living condition of an individual while assessing the impact of the disease on the impairment of daily activities, of emotional, functional and psychosocial states and life prospects [20]. It is something that is personal and can only be self-measured [21]. Thus, this study aimed to evaluate the quality of life of sickle cell disease carrier's in the micro-region of Guanambi, State of Bahia, treated under the Sickle Cell Anemia Program (PAF).

Method

This is a quantitative-descriptive-analytical study conducted in the micro-region of Guanambi, State of Bahia, including five of the eighteen municipalities that are part of the study area, namely Guanambi, Iuiú, Riacho de Santana, Palmas de Monte Alto and Pindaí. The survey worked with one case group and one control group, where the case group was represented by sickle cell disease carriers treated under the SCDP-Sickle Cell Disease Program, aged 12 years and over, regardless of sex. The control group included people who were free from any chronic disease that could directly affect the quality of life, but with socio-demographic and economic features similar to case group participants, namely, regarding age, family income and municipality of origin.

Quality of life-assessment tools SF-36 (Medical Outcomes Study 36-Item Short-Form Health Survey)-and WHOQOL-BREF (tool developed by the World Health Organization) were used for data collection, as well as an economic and socio-demographic questionnaire prepared by researchers. Fisher's exact test was applied to compare the socioeconomic and demographic variables in both groups. Before choosing the mean comparison test, data were analyzed with the Kolmogorov-Smirnov test to verify whether they followed normal distribution. Student t and Mann-Whitney tests were used to compare income and age of SCD (spelled) and non-SCD participants. The Student t test was applied to verify the probability of significant difference between the mean scores of the two tools (SF-36 and WHOQOL-BREF) in the two groups. Spearman correlation evaluated the association between the scores of the two tools and correlated with age.

The study was conducted in conformity with ethical principles according to Resolution 466/12, with the approval of the Ethics in Research Committee (CEP) under opinion number 889.040.

Results

A total of 20 SCD were they all carrier's sickle cell anemia? Specify the characteristics of this group; if possible insert this data in Table1 and 40 non-SCD participants were evaluated in this study. Women prevailed in both groups. SCD carriers had lower schooling than the healthy group (p=0.0202) and black people were more frequent in the SCD-affected group than in the SCD-free group (p=0.0135) (Table 1).

Table 1: Sociodemographic characteristics of SCD and Non-SCD participants, Guanambi, State of Bahia (2015).

Variables	SCD	Non-SCD	*p**
	n (%)	n (%)	
Gender			
Men	9 (45)	9 (22)	0.1335
Women	11 (55)	31 (78)	
Schooling			
With schooling	17 (85)	40 (100)	0.0202
Without schooling	3 (15)	0	
Own home			
Yes	17 (85)	30 (75)	0.5128
No	3 (15)	10 (25)	
Skin color			
Black	8 (40)	4 (10)	0.0135
Other	12 (60)	36 (90)	

*Fisher's Exact Test
SCD: Sickle Cell Disease.

Table 2: Comparison of SF-36 mean components scores between SCD (n=20) and Non-SCD patients (n=40), Guanambi, State of Bahia (2015).

SF-36 Components	SCD (n=20)	Non-SCD (n=40)	*p**
	Mean (±sd)	Mean (±sd)	
Functional capacity	40.50 (±17.98)	80.12 (±19.67)	< 0.0001
Physical aspects	46.25 (±39.96)	82.50 (±27.85)	0.0001
Pain	51.30 (±27.58)	67.20 (±26.91)	0.0365
General state of health	53.35 (±19.59)	58.23 (±17.05)	0.3247
Vitality	51.75 (±21.72)	68.00 (±19.28)	0.0045
Social aspect	76.88 (±21.94)	75.31 (±24.92)	0.8128
Emotional aspect	38.32 (±40.86)	68.32 (±38.46)	0.0071
Mental health	53.00 (±24.96)	68.00 (±22.52)	0.0223

*Student t test, sd= Standard Deviation
SCD: Sickle Cell Disease not needs to repeat it all time. The first time is sufficient to all the text.

The mean age of groups studied was 30 years and the median family income was R$ 788.00. The general state of health and the social aspect did not differ between SCD carriers and non-SCD individuals. However, patients with SCD showed lower quality of life compared with non-SCD people when observing the components of functional capacity (p<0.0001); physical aspects (p=0.0001); pain (p=0.0365); vitality (p=0.0045); emotional aspect (p=0.0071); mental health (p=0.0223) (Table 2). Both emotional aspect and functional capacity were the lowest scoring components in SCD patients.

Of the SF-36 tools components, physical aspect had a negative correlation with age (p=0.0238), indicating worse general physical condition with age (Table 3). Mean sickle cell patient age was 32.7 (dp±18.8).

Using the WHOQOL-BREF instrument that evaluated physical,

Table 3: Correlation between SF-36 components and mean age of SCD patients, Guanambi, State of Bahia (2015).

SF-36 Components	Age	
	R	*p**
Functional capacity	-0.1879	0.4276
Physical aspects	-0.5029	0.0238
Pain	0.0688	0.7731
General state of health	-0.3169	0.1733
Vitality	-0.3914	0.0878
Social aspect	-0.2055	0.3848
Emotional aspect	-0.3175	0.1725
Mental health	-0.1171	0.6229

Table 4: Comparison of mean scores of WHOQOL-BREF components between patients with and without SCD, Guanambi, State of Bahia (2015).

WHOQOL-BREF Components	SCD (n=20)	Non-SCD (n=40)	*p**
	Mean (±sd)	Mean (±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
Environmental	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 t test, sd= Standard Deviation
SCD: Sickle Cell Disease

Table 5: Correlation between the SF-36 and WHOQOL-BREF components among patients with Sickle Cell Disease (SCD), Guanambi, State of Bahia (2015).

SF-36 Components	WHOQOL-BREF Components		
	Physical	Psychological	Social
Physical aspects	0.5586*	---	---
Mental health	---	0.6773*	---
Social aspects	---	---	0.5484*

*Spearman's Correlation (*p*)

psychological, social, environmental and overall quality of life components, only physical aspects and the overall quality of life had lower scores in the SCD carrier group ($p<0.05$) (Table 4). However, none of the WHOQOL-BREF components had a correlation with the age of SCD carrier group participants.

No significant associations were found between the components of the two tools (Table 5).

Discussion

Our results have demonstrated that the quality of life of SCD carriers is perceived by them as poor, especially when related to physical and social aspects. This study also demonstrated the need to apply more than one tool in the assessment of quality of life, since no correlation was found between them.

The evaluated population group is historically less favored because it is a region with the largest number of African descent people and unfavorable socioeconomic situation, which may reflect in low education level. Another factor that may be influencing low schooling level of the assessed group are the disease-imposed constraints, preventing or hindering access by SCD carriers to formal education [15].

Poor schooling is linked to the high rate of misinformation about the disease. The lack of knowledge about sickle cell disease limits the participation of SCD carriers in social groups, such as the school environment, hampering the development of their personality and consolidation of their social relationships [22].

The general health component analyzes the patient's perception regarding his overall health [23]. Although few studies address the psychosocial issue in people with sickle cell disease (SCD), Figueiredo [24], Felix, and Souza Ribeiro [25] say that there are major issues related to chronic disease with regard to social aspects, such as problems in family relationships, interaction with colleagues and friends and in academic performance. Socioeconomic problems may escalate psychological problems, such as difficulties to relate emotionally, pegged to a low self-esteem [15]. However, in our study, SCD carrier participants showed interaction with social activities. According to Figueiredo [24] ref, even in global terms, unless we understand their living environment, schooling conditions, work and social life in general, we shall but have only a small glimpse of the disease and hence currently proposed solutions to improve the situation shall have little effectiveness.

Functional capacity refers to the individual's ability to perform basic physical and mental daily activities [26]. When assessing the functional capacity of participants with SCD, index was considered bad. This may result from constant vaso-occlusion processes that can lead to complications in various organs which, and these changes decrease the functional capacity of SCD carriers, especially those patients with sickle cell anemia [27].

SCD affects the physical condition of the patient, possibly due to the symptoms of the disease itself, showing treatment dependency and proneness to fatigue and pain [28]. Pain is one of the most prevalent medical conditions in sickle cell disease, it is marked by the presence of painful crises, paleness, tiredness, jaundice, leg ulcers and increased likelihood of infection [29].

Santos *et al.,* [30] say that pain has great negative impact on the quality of life of patients with SCD and even their families, and their evaluation is very useful in clinical practice to demonstrate the extent of the problem. A study by O'Hara *et al.*, [31] pointed out that, because of the pain in the body regions analyzed, it was found that some areas concerning the SF-36 physical component (functional capacity, pain, physical aspects and general state of health) evidenced significant reduction. This shows that constraint due to physical aspects can be directly affected due to pain endured by the patient.

The vitality component (SF-36) addresses issues that consider the patient's energy and fatigue levels. Patients with chronic disease often experience changes in this area as a result of pain symptoms, physical and psychological aspects that cause unwillingness to perform daily activities [32].

The limitation due to the emotional aspect, which enables the assessment of the impact of psychological aspects in an individual's well-being, achieved the worst mean among SF-36 components, showing that this factor has a major impact on the quality of life of SCD patients. In their study, O'Hara *et al.,* [31] say that compromised social aspects and limitation due to emotional aspect components do not seem to be associated with pain felt by patients, but rather are influenced by other components of the disease. The instability caused by the disease, linked to often unfavorable socioeconomic profile thereof may explain such impact.

The mental health component addresses issues about state

of mind, mood, anxiety and psychological well-being. Disease complications can lead to psychological disorders such as depression, with impairment in the quality of life when associated with symptoms of this disorder [33].

With age, it is common that some factors arise and affect the quality of life, such as instability, vulnerability to disease-imposed constraints, intellectual difficulty, including gradual loss of own role in society hitherto performed. These factors adversely influence the quality of life, making the individual more dissatisfied with his own condition [34,35].

Tool choice was one of the most important points related to the purpose of the study. Tools had to be linked to a cultural context compatible to the surveyed population [36]. Due to specific elements, this study used both instruments to try to minimize their important limitations by proposing indicators that fail to assess the specificity of each subject in each assessed context [37]. WHOQOL-BREF is based on the concept of Quality Of Life (QOL) of WHO, addressing aspects that encompass comprehensively QOL, including factors that are related and unrelated to health, in other words, it evaluates the general quality of life. SF-36 was developed to assess health aspects and activities generally affected by health conditions, evaluating only health-related quality of life, in other words, it is a more health-specific QOL tool. Therefore, one can observe very weak or no significant correlations between the components of these two questionnaires [36], as shown in this study [38].

The study's major limitation was the reduced sample size, which only allows considering the results for the population at hand.

Conclusion Please, Add Few More Proposition or Solution to Ameliorate Quality of Life of SCD in this Region

The main points highlighted by the study were that patients with sickle cell disease suffer major negative impact on the quality of life; they are mainly black and have a lower education level.

The emotional and physical aspect and the functional capacity were the SF-36 and WHOQOL-BREF components with worse scores in SCD patients. The physical aspect is compromised with age.

The linkage of the two tools used in the study evidenced no correlation between them. Tool choice should direct the desired study toward evaluating quality of life related to health or in general, since tools used here show differences when tested to health aspects-oriented quality of life.

The research points to the importance of analyzing the quality of life, showing that it interferes and is also influenced by the health of the patient with sickle cell disease.

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