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Evaluation of quality of life of patients with sickle cell disease in a General Hospital of Goiás, Brazil

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Introduction: Sickle cell disease is the most common inherited disease in Brazil. Patients are known to suffer physical, emotional and social impairment and their quality of life may well be involved. *Method:* The quality of life of sickle cell disease patients treated in Hospital das Clínicas of the Universidade Federal de Goiás was evaluated. Sixty patients with ages ranging from 14 to 60 years old were interviewed. The WHOQOL-Bref (a quality of life validation instrument of the World Health Organization), and the ethnical-racial, and sociodemographic questionnaires were administered. A standard error of 5% ($p\text{-value} \leq 0.05$) was considered acceptable. *Results:* The mean age of the participants was 27 years old, 53.3% of the patients were women, 71.7% were single and 51.7% had completed elementary school. The majority classified themselves as mulattos (46.7%) and the minority Blacks (11.7%). Only 6.7% considered themselves victims of racial discrimination because of their skin color but 33.3% considered themselves victims of discrimination due to sickle cell disease. The patients 48.3% reported an association between their disease and their skin color. The quality of life was considered bad by 6.7% and good by 70%. A total of 48.3% considered their lives to be satisfactory and 23.3% to be unsatisfactory. The scores obtained from the WHOQOL-Bref (from 0 to 100) were: 57.32 for physical, 66.03 for psychological, 69.86 for social and 52.76 for environmental domains. There were significant correlations of discrimination due to the disease with educational level and age with all the WHOQOL-Bref domains. *Conclusion:* Sickle cell disease significantly limits the quality of life of patients. Also, sickle cell disease, coinciding with the racial miscegenation, is losing its "black-related disease" character in Brazil.

Keywords: Disease, sickle cell; chronic disease; Quality of Life

Introduction

Sickle cell disease (SCD) is a hereditary disease caused by a mutation that leads to the substitution of glutamic acid by valine at the sixth position in the β -globin chain. The genetic anomaly produces changes in the hemoglobin

molecule so that, at low oxygen tensions, these molecules undergo a polymerization process and lose the deformability characteristics of red blood cells, leaving them with a "sickle" shape. These sickled red blood cells aggregate in capillaries, causing partial or total obstruction of the vessel lumen.⁽¹⁾

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The clinical signs and symptoms are related to severe anemia and chronic hyperbilirubinemia and, in particular, vaso-occlusive complications including vaso-occlusive crises.⁽¹⁾

The estimated prevalence of individuals with sickle cell trait (heterozygotes) in the Brazilian general population is 4%, that is, currently 7.2 million cases are carriers, with an estimation of 200,000 new cases annually.⁽²⁾ Additionally, it is estimated that there are 25 to 30 thousand homozygotes in Brazil with 3500 new cases born each year.⁽²⁾ In Goiás, the estimated proportion of individuals carrying the sickle cell trait is 2.2%.⁽³⁾

In Brazil the prevalence is still higher among blacks, however SCD is distributed heterogeneously and involves white individuals due to the miscegenation of the Brazilian population.⁽⁴⁾

Thus, SCD is the most common monogenic hereditary disease in Brazil coursing with much clinical variability, from almost asymptomatic patients to those with many complications and frequent hospitalization. Hence this disease is considered a public health problem.

Despite the advances in medicine, this disease remains incurable and only palliative care can be provided. It is therefore a chronic disease with lifelong treatment with the quality of life (QoL) of patients being a key challenge of patients, their families and healthcare professionals.⁽⁴⁾

According to the World Health Organization (WHO), QoL is defined as the "Individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns."⁽⁵⁾

Moreover, there is the concept of "quality of life associated with health," which assesses the impact of health on the individual's capacity to live fully.⁽⁶⁾

Over the last thirty years, the assessment of QoL has gradually become important in healthcare, with improvements and sophistication in evaluation methods focusing on subjective evaluations to measure individual perceptions of QoL of patients.⁽⁷⁾

According to Padilla,⁽⁸⁾ the concept of QoL related to health has gained a multidimensional nature with physical, psychological, social and spiritual well-being; these last three dimensions are often more valued than the physical dimension.

Due to the importance of QoL in the context of the evolution and treatment of diseases, especially chronic diseases, this variable has been included in clinical trials as an aspect to be evaluated with the aim of valuing broader parameters than just the control of symptoms, decreased mortality and an increase in life expectancy.⁽⁹⁾

According to Tapper,⁽¹⁰⁾ the first cases of SCD diagnosed in the early twentieth century, highlighted a close relationship between race and disease, with this disease assuming the character of a "black-related disease". Even today, despite the compensatory racial policies and changes in social relationships, SCD is often seen by patients as a stigma.

It is in this scenario that an attitude emerges that sees the illness as a punishment, thereby aggravating the patient's perception regarding their exclusion. And in the case of SCD, the punishment is twofold: both having the illness and due to the ethnicity.

Allied to the issue of race are precarious social and economic conditions, which is a significant risk factor for depression. In addition, personal (personality and individual perception about the disease, among others) and family (cohesion and family support, for example) factors strongly influence a possible depressive condition.⁽¹⁰⁾ Individuals who lack adequate social, economic and family support tend to have more problems associated with the disease and more difficulties to adjust to it. This may be the trigger of painful episodes, which have been associated with high levels of depressive symptoms.⁽¹¹⁾

The importance to expand discussions involving the QoL of SCD patients becomes apparent; it is a chronic disease that is deemed a public health problem in Brazil. The consequences in patients' lives are numerous and include certain limitations. Understanding these limitations allows us to identify problems and devise appropriate measures to intervene and modify variables that negatively affect QoL.⁽⁹⁾

Thus, in agreement with the organized social segments of black men and women in Brazil, we proposed to investigate and analyze indicators of the QoL of SCD patients under treatment.

We evaluated the QoL through physical, social, psychological and environmental indicators using the World Health Organization QoL assessment instrument - Abbreviated (WHOQOL-Bref) and compared the data obtained from employing demographic and ethnic-racial questionnaires.

Method

The research was conducted using quantitative resources producing a descriptive, exploratory and cross-sectional study.

Sixty consecutive SCD patients who went to the Hemoglobinopathy Clinic of the Hematology Service in the period from January to July 2008 were interviewed from a population of 120 male and female SCD patients under treatment. The ages of the participants varied between 14 and 60 years old. The sample is representative of the general population with a margin of error of 5%.

Patients or guardians were informed about nature of the research and that participation was voluntary before they gave their informed written consent. Under 14-year-old and over 60-year-old patients were excluded.

The WHOQOL-Bref was derived from another instrument called the World Health Organization QoL Assessment 100 (WHOQOL 100), both of which were validated in Portuguese by Fleck et al.^(12,13) While other QoL

assessment instruments have been developed, it appears that, to date, this one has the highest value in research.⁽⁹⁾

WHOQOL-Bref consists of 26 questions, two general and 24 representing each of the 24 aspects that make up the original instrument. Thus, unlike the WHOQOL-100, in which each of the 24 aspects is assessed based on four key questions, in WHOQOL-Bref each aspect is assessed by a single question.

The first two questions of the WHOQOL-Bref relate to the individual's perception of the QoL, and the individual's perception of health.

The remaining 24 questions comprise four domains: physical, psychological, social and environmental.

The physical domain is investigated by questions 3, 4, 10, 15, 16, 17 and 18, the psychological domain by 5, 6, 7, 11, 19 and 26, the social domain by questions 20, 21 and 22 and the environmental domain by 8, 9, 12, 13, 14, 23, 24 and 25.

All questions are presented as a Likert scale of five points from 1 (very bad) to 5 (very good QoL). To score the WHOQO-BREF, the questions are separated by domain and the average of all participants is calculated, i.e. the scores of the questions of each domain are summed and divided by the total number of participants. The scores of each domain can then be converted to a 0-100 scale.

Asnani et al. demonstrated the effectiveness of the WHOQOL-Bref to measure the QoL of SCD patients, underscoring the fact that this instrument was developed from scattered groups in research centers worldwide with the goal of creating a questionnaire that could be used to measure the QoL of diverse populations in different countries.⁽¹⁴⁾

The ethnic-racial questionnaire is an instrument created by the authors of this paper to detect whether or not there is a relationship between the patient's illness and ethnic background. It consists of four questions, the first on the participant's opinion as to his/her ethnic background based on the ethnic classification system of the Brazilian Institute of Geography and Statistics (IBGE). The second and third relate to discrimination in relation to the skin color and disease, respectively and the last question is related to discrimination related to the ethnic background and the disease to see if the patient believes his/her illness is or is not related to the skin color.

The sociodemographic questionnaire is an instrument created by the authors that collected data on gender, age, schooling and marital status.

After presenting the research project of the health team of the Hematology Department and approval of the Research Ethics Committee, recruitment of potential research subjects was started through direct contact during medical consultations. The objectives of the investigation and the data collection procedures were explained. If patients agreed they signed two informed consent forms; one remained with the researcher and one with the patient. None of the

consecutive 60 potential subjects that were asked refused to participate.

The data collection process occurred concurrently to the medical consultation. During the consultation the WHOQOL-Bref ethnic-racial and sociodemographic questionnaires were applied by the researchers.

The results of the WHOQOL-Bref were assessed according to the scores obtained for each domain, after conversion to a 0-100 scale, using the SPSS® version 16.0 computer program with specific syntax for the WHOQOL-Bref. The results of the WHOQOL-Bref were then compared with demographic data obtained by the socioeconomic questionnaire.

The results of the ethnic-racial questionnaire were evaluated according to the frequency of answers for each question. These results were then compared to demographic data. The level of significance for all tests was set at 5% ($p < 0.05$).

Results

The sociodemographic questionnaire (Table 1) revealed that most subjects were female (53.3%) and unmarried (71.7%). The mean age was 27 years old ranging from 14 to 60 years and the educational level was up to eighth grade (51.7%).

According to the ethnic-racial questionnaire, 46.7% of individuals considered themselves mulattos, 13.3% considered themselves white and 11.7% considered themselves black and the other 28.3% of the participants preferred not answer this question. A total of 6.7% of the participants said they were discriminated against because of

Table 1. Frequency (n) and percentages (%) of sociodemographic data of the 60 sickle cell disease patients under treatment

Variables	n	(%)
Gender		
Male	28	(46.70)
Female	32	(53.30)
Age		
14 - 30	40	(66.7)
31 - 45	15	(25.0)
46 - 60	5	(8.3)
Marital status		
Single	43	(71.7)
Married	9	(15.0)
Live-in partner	5	(8.3)
Separated	0	(0)
Divorced	2	(3.3)
Widowed	1	(1.7)
Schooling		
Illiterate	2	(3.3)
Elementary school incomplete	25	(41.7)
Elementary school completed	4	(6.7)
Junior high school incomplete	7	(11.7)
Junior high school completed	19	(31.7)
Senior high school incomplete	2	(3.3)
Senior high school completed	1	(1.6)

their color, while 33.3% said they were victims of prejudice about the disease. Additionally, a total of 48.3% said that there is a relationship between their illness and color, while 50.0% do not believe this relationship exists. The other 1.7% of the participants preferred not to answer this question.

Those who made up the 14- to 30-year-old age group did not believe there was a link between the illness and color a result that was different to individuals who were over 30 years old.

On assessing the replies to question 1 of the WHOQOL-Bref instrument, 6.7% rated their QoL negatively, that is bad and very bad, 23.3% rated it as neither good nor bad and 70.0% evaluated it as good or very good.

In respect to question 2 of the WHOQOL-Bref, 23.3% were dissatisfied or very dissatisfied with their health, 28.4% were neither satisfied nor dissatisfied and 48.3% were satisfied or very pleased.

Although the relationship between health and QoL was recognized many years ago, it was realized that the perceptions of individuals regarding their QoL was more positive than their perceptions about their health state ($p < 0.05$).

The scores of the WHOQOL-Bref as 0-100 scales were as follows: physical = 57.32, psychological = 66.03, social = 69.86 and environmental = 52.76.

There was a significant correlation between discrimination due to illness and educational level (Table 2) and between age and all domains of the WHOQOL-Bref instrument (Table 3).

Table 2. Correlations between the variables on the sociodemographic and ethnic-racial questionnaires

Variables	Coefficients	Gender	Age	Schooling	Marital status
Color	r	-0.036	-0.086	0.035	-0.137
	p-value	0.784	0.511	0.789	0.296
Discrimination due to color	r	0.113	0.101	-0.040	0.053
	p-value	0.389	0.403	0.764	0.689
Discrimination with disease	r	0.209	0.031	-0.324	0.160
	p-value	0.108	0.812	0.012	0.223
Disease linked to color	r	-0.067	-0.204	-0.183	0.086
	p-value	0.611	0.118	0.162	0.514

Pearson coefficient = r, significance: p-value \leq 0.05

Table 3. Correlations between the domains of the WHOQOL-Bref and demographic data

Variables	Coefficients	Physical domain	Psychological domain	Social domain	Environmental domain
Gender	r	0.046	-0.114	-0.051	-0.105
	p-value	0.728	0.385	0.701	0.425
Age	r	-0.398	-0.368	-0.285	-0.33
	p-value	0.002	0.004	0.027	0.01
Schooling	r	-0.109	0.115	-0.046	-0.162
	p-value	0.407	0.236	0.729	0.216
Marital status	r	-0.035	-0.163	-0.132	-0.129
	p-value	0.788	0.213	0.315	0.326

Pearson coefficient = r, significance: p-value \leq 0.05

Discussion

Chronic diseases in general create physical, emotional and social alterations that demand coping mechanisms which often have a direct impact on the QoL.

SCD, as a chronic disease, causes such changes. These changes develop from various complications that arise during the natural course of the disease. Additionally the life expectancy of the individual is reduced. In this investigation, we noted that the results are correlated to the findings of the study by Alves on mortality due to SCD.⁽¹⁵⁾

The data show that 66.7% of the sample was 30 years old or younger, similar to the results of Alves, who found that 70% of SCD patients did not survive past 30 years old.⁽¹⁵⁾

The Pain in Sickle Cell Epidemiology Study (PiSCES) compared the QoL of SCD patients with the QoL of patients with other chronic diseases, such as asthma, and hemodialysis patients and found that they were similar.⁽¹⁶⁾

There is also a close relation of the disease with black people; these individuals suffer from two stigmatizing features, the presence of SCD and the ethnic origin.

We believe that, although SCD is overcoming the character of being a "black-related disease" because of intense racial miscegenation in Brazil,⁽¹⁰⁾ there is still an association between clinical data and prevalence among African descendants. These socially marginalized individuals thus suffer a significant loss in QoL.

In an analysis of the demographic data, it was found that the participants are still in the economically active age with most (66.7%) being between 14 and 30 years old. In a study of Asnani et al., the mean life expectancy for SCD patients was 53 years for men and 58.5 years for women. (14) Another study found that about 11% of patients with SCD do not survive to adulthood.⁽¹⁷⁾ Thus, can one infer that low QoL worsens the patient's life expectancy?⁽¹⁸⁾

The ethnic-racial questionnaire showed that 46.7% of individuals considered themselves mulattos, 13.3% considered themselves white and 11.7% considered themselves black and the other 28.3% of the participants did not answer this question. Knowing the close relationship between race and SCD,⁽¹⁰⁾ a higher proportion of Blacks was expected. The result may reflect the ethnic heterogeneity of the Brazilian population⁽¹⁹⁾ or the fear of being characterized as black.

Another analysis was about discrimination related to color and disease: 6.7% of participants said they were discriminated against in regards to color, while 33.3% said they were victims of discrimination because of their disease, showing that disease can have a greater negative influence on social relations than color. When asked about their impression about the link between SCD and color, 48.3% responded that the relationship is valid as against 50.0% who do not believe this relationship exists. Moreover, it was observed that 14 to 30 year-old individuals did not believe that this link between illness and color existed while over 30

year olds did. This finding indicates that older individuals are more aware about their disease than younger ones, suggesting a greater interest in learning about the disease and how it may affect their QoL.

By analyzing questions 1 and 2 of the WHOQOL-Bref on general issues about QoL and health, it was found that although sick, most of the individuals positively evaluated both their QoL and their health.

Although the relationship between health and QoL was recognized several years ago,⁽²⁰⁾ it is important to realize that the individuals' perceptions of their QoL were more positive than their perceptions about their health status ($p < 0.05$).

On analyzing the four domains (Physical, psychological, social and environmental), scores were calculated for each domain on a scale of 0 to 100. The most significant loss of QoL according to the results of the WHOQOL-Bref was the environmental domain with a mean score of 52.76%, followed by the physical domain with a mean of 57.32%.

Pereira et al. found significant decreases in the physical, psychological and social domains for SCD patients.⁽⁴⁾ The association with the physical domain should be noted; the disease affects the physical domain as the patient is dependent on medical treatment and is more susceptible to fatigue and pain, and suffers reduced working capacity.

The environmental domain was the most affected area; this may be because the study population was from a government hospital where the vast majority is treated under the conditions of the public health system. Probably these are low-income subjects with poor housing and few options of leisure activities.

On analyzing the physical domain, the score decreased with advancing age probably due to normal tiring of individuals as they grow old or due to worsening symptomatology of the disease or even a combination of these two factors.

When the psychological domain is analyzed, it seems that older individuals have more psychological problems such as low self-esteem and worry about death.

The social domain score had the highest scores indicating that SCD individuals have social support, i.e. individuals trained to deal with environmental stressors.

There is also a possible negative correlation (negative r) between age and all domains of the WHOQOL-Bref (Table 3). A negative value for r means that the variables compared go in opposite directions, i.e. if one increases the other decreases and vice versa. Thus, the negative correlation in this case indicates that as age increases the individual's domain scores diminish.

Another possible correlation is that of prejudice against the disease and the schooling of participants (Table 2), the case of a negative correlation indicates that as the number of years of schooling increases, the perception of discrimination against the disease decreases.

Conclusion

Data from this study confirm that SCD, as it is a chronic disease and courses with great variability, causes limitations in the lives of sufferers, with the pain and hospitalization being most likely responsible for the destabilization of the physical and emotional aspects of the individual.

It was also found that the disease is losing its nature as a "black-related disease", coinciding with Brazilian racial miscegenation.

An important aspect to be addressed by future studies is to compare the QoL of subgroups of SCD patients to assess whether the association with other hemoglobinopathies and therapeutic interventions affects the QoL of patients. In this work, in particular, one can say that there was loss of QoL of SCD patients under treatment.

QoL is an important aspect to be analyzed, especially as it is multidimensional, influencing and being influenced by the health of the individual.

In addition to treating pain, efforts to improve the QoL of SCD patients should incorporate the psychological aspect, aimed at reducing anxiety, depression and family problems.⁽²¹⁾

In the case of chronic diseases, QoL gains greater importance as it directly influences the prognosis of the disease and so more research is needed on the subject, in order to formulate strategies of healthcare programs and other measures to care for patients.⁽⁹⁾

Resumo

Introdução: A doença falciforme é a doença hereditária mais frequente no nosso país. O portador apresenta acometimentos físico, emocional e social, e sua qualidade de vida pode estar comprometida. Objetivo: Avaliar a qualidade de vida dos doentes falciformes em tratamento no Hospital das Clínicas da Universidade de Goiás. Método: Foram entrevistados 60 sujeitos entre 14 e 60 anos, doentes falciformes, em tratamento no Hospital das Clínicas. Aplicou-se o WHOQOL-Bref (instrumento avaliativo de qualidade de vida da Organização Mundial de Saúde – OMS), o questionário étnico-racial e o sociodemográfico. A significância foi definida por um erro padrão de 5% ($p \leq 0,05$). Os sujeitos eram do sexo feminino em 53,3% e solteiros em 71,7%. A média da idade foi de 27 anos e o nível educacional até o primeiro grau completo foi de 51,7%. Resultados: A maioria considerou-se parda (46,7%) e a minoria, negra (11,7%). Apenas 6,7% disseram ser vítimas de preconceito devido à cor e 33,3% disseram ser vítimas de preconceito devido à doença. Os sujeitos relataram ligação entre doença e sua cor em 48,3%. A qualidade de vida foi avaliada negativa em 6,7% e, em 70%, positiva. Apresentaram satisfação negativa quanto à saúde 23,3% dos sujeitos e, em 48,3%, a satisfação foi positiva. Os escores do WHOQOL-Bref, de 0 a 100 foram: domínio físico (57,32), psicológico (66,03), social (69,86) e ambiental (52,76). Conclusão: Houve correlação significativa entre preconceito devido à doença e nível educacional, e entre idade e todos os domínios. A doença falciforme limita a vida do portador, com comprometimento da

qualidade de vida. A doença está perdendo o caráter de "black related disease", coincidindo com a miscigenação racial brasileira.

Descritores: Anemia falciforme; Doença crônica; Qualidade de vida; Questionários; Fatores socioeconômicos; Grupos étnicos

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