

Sociodemographic aspects and quality of life of patients with sickle cell anemia

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Background: Sickle cell anemia is a chronic inherited disease, widespread in the Brazilian population due to the high degree of miscegenation in the country. Despite the high prevalence, there are few studies describing the characteristics of patients and the impact of the disease on quality of life.

Objective: To describe the sociodemographic profile and the impact of the disease on the quality of life of sickle cell anemia patients.

Methods: Over 18-year-old patients with sickle cell anemia who attended meetings held by the Associação Baiana de Portadores de Doenças Falciformes, an association for sickle cell anemia patients in Bahia, were interviewed. Sociodemographic data were collected and the generic the Medical Outcomes 36-Item Short-Form Health Survey (SF-36) questionnaire, which is used to assess quality of life, was applied. The analysis of the descriptive statistics was performed using the Statistics Program for the Social Sciences software.

Results: Thirty-two mostly female (65.6%) patients were interviewed. The mean age was 31.9 ± 12.67 years, 50.0% considered themselves black, 68.8% did not work and 87.5% had per capita income below the poverty line (up to one and a half minimum wages). The SF-36 scores were: limitation by physical aspects 26.56, functional capacity 28.9, emotional aspects 30.20, social aspects, 50.0, pain 50.31, mental health 54.62, general health status 56.09 and vitality 56.71. This shows that the disease has a huge impact on the patients' quality of life.

Conclusion: The disease interferes in the working capacity of individuals, who mostly have low incomes and impaired access to healthcare services and significantly impacts on their quality of life.

Keywords: Anemia, sickle cell; Chronic disease; Quality of life; Health profile; Questionnaires; Socioeconomic factors

Introduction

Sickle cell anemia, described for the first time by James Herrick in 1910⁽¹⁾, is the most common inherited hematological disease in the world⁽²⁾. It was initially identified in Africa and, although it does not only affect individuals of African origins⁽¹⁾, the prevalence is higher in black populations and their descendants. In Brazil, between 0.1 and 0.3% of black population is affected^(2,3). The disease affects 6.3% of the afro-descendant population in Bahia; a study involving afro-descendant children in the city of Salvador demonstrated a frequency of 7.4%⁽⁴⁾. Sickle cell anemia is described as an incurable genetic disease which, although treatable, generally causes a high level of suffering to its carriers and, for this reason, deserves special attention from the medical, genetic and psychosocial perspectives⁽⁵⁻⁷⁾.

The cause of this disease is a genetic mutation at position 6 of beta chain of hemoglobin, the component of red blood cells that is responsible for transporting gases in the blood⁽⁸⁾. A distortion of the shape of red blood cells, which start presenting a sickle shape and become more rigid, can cause vaso-occlusive complications, reduced oxygenation, progressive tissue dehydration and reductions in organic functions^(9,10).

People with sickle cell anemia present several signs and symptoms that can occur in different ways and at different severities; they occur jointly or separately. These include chronic anemia, painful crises, infection, fever, jaundice, splenic sequestration (retention of blood in the spleen), leg ulcerations, priapism and stroke, amongst others⁽¹¹⁾. These complications have a physical, emotional and social impact on the carrier's life that can compromise their quality of life (QOL).

World Health Organization (WHO) defines QOL as "an 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". Recognition of the multidimensional aspects involved intends to evaluate the effect of certain events and acquisitions in people's life such as health status, cognitive function, sexual satisfaction, daily activities, emotional well-being and family and social life⁽¹²⁾.

Although QOL has been studied in several social groups, it is not widely known in patients with sickle cell anemia, especially in Brazil, where studies on sociodemographic aspects are still scarce. Knowledge of these data allows the identification of and approach to these patients' needs regarding improving their QOL, and would contribute to improve

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resource allocation and to create public health policies to benefit this population. That said, the aim of this study was to characterize the sociodemographic aspects and QOL of people with sickle cell anemia.

Methods

A cross-sectional descriptive study was performed using a quantitative approach at an association for sickle cell anemia patients in Bahia (Associação Baiana de Portadores de Doenças Falciformes - ABADFAL) located in the City of Salvador. All over 18-year-old patients diagnosed by a specialist as having sickle cell anemia who attended the meetings organized by ABADFAL during the period between October 2010 and May 2011 were invited to participate in the study.

All the individuals were instructed about the research procedures and objectives and those who agreed to participate in the study were required to sign an informed consent form. This study had previously been approved by the Ethics Research Committee of the University of Salvador (# 04.11.46) according to Resolution 196/96 of the Conselho Nacional de Saúde.

At first the authors elaborated and applied an evaluation questionnaire to collect sociodemographic data, such as gender, age, ethnic background, schooling, marital status, work (employed or unemployed) and per capita income.

Secondly, a standardized QOL questionnaire, the Medical Outcomes 36-Item Short-Form Health Survey, as adapted and validated in Brazil by Ciconelli et al., was applied⁽¹³⁾. This questionnaire is composed of 36 questions to assess QOL by specific domains: functional ability, physical aspects, pain, general health perception, vitality, social and emotional aspects and mental health. It uses a score of 0 to 100, in which 0 corresponds to the worse general health status and 100 corresponds to the best general health status⁽¹³⁾.

Data were collected in single individual interviews and after collection they were digitized using the Excel® version XP software. Descriptive statistics was used for the analysis of sociodemographic and QOL data; continuous variables were analyzed as central tendency measures and were expressed as means and standard deviations. The categorical (dichotomous) variables were analyzed as measures of frequency and expressed as percentages. A sub-analysis was performed with stratification of the patients in subgroups in respect to work (employed or unemployed) and schooling (incomplete secondary education, complete secondary education and higher education). The Student t-test for independent samples was used to make this comparison. Statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS) software for Windows (version 14.0).

Results

Thirty-two adult patients were interviewed; most of them were women (65.6%) with a mean age 31.90 ± 12.67 years (range: 18-55 years). The majority were Black with complete secondary education, unemployed and had per capita income of up to one and a half minimum wages (87.5%). The sociodemographic characteristics are described in Table 1.

Table 1 - Distribution of sociodemographic variables obtained from the sample of 32 patients attending the Associação Baiana de Portadores de Doenças Falciformes

	n	%
Gender		
Female	21	65.6
Male	11	34.4
Skin color		
White	2	6.3
Black	16	50.0
Mixed	13	40.6
Indigenous	1	3.1
Schooling		
Incomplete primary school	7	21.9
Complete primary school	2	6.3
Incomplete secondary education	10	31.3
Complete secondary education	11	34.4
Incomplete higher education	1	3.1
Complete higher education	1	3.1
Marital status		
Single	21	65.6
Married	7	21.9
Common law marriage	1	3.1
Separated	3	9.4
Employment		
remunerated	6	18.8
Volunteer	4	12.5
Unemployed	22	68.8
Income		
Up to 1.5 MW	28	87.5
From 1.5 to 3 MW	3	9.4
From 3 to 5 MW	1	3.1

MW: minimum wage

The mean scores for the different domains as assessed by SF-36 are presented in Table 2. An overall reduction in QOL domain scores was observed, which demonstrates the negative impact of the disease on QOL.

Table 2 - Results of values obtained using the Medical Outcomes 36-Item Short-Form Health Survey applied to 32 patients with sickle cell anemia

Domain	Mean \pm SD	Minimum	Maximum
Functional ability	28.9 \pm 16.69	0	50.0
Physical limitations	26.56 \pm 29.05	0	100.0
Pain	50.31 \pm 17.86	0	80.0
General health status	56.09 \pm 12.22	35.0	80.0
Vitality	56.7 \pm 13.29	30.0	80.0
Social aspects	50.0 \pm 17.96	25.0	87.5
Emotional aspects	30.20 \pm 33.18	0	100.0
Mental health	54.62 \pm 12.71	24.0	72.0

The stratification of patients in respect to work showed that 68.8% were unemployed. A comparison between QOL domains among those who worked and those who did not, did not give statistically significant differences for any of the domains (p-value > 0.05). The same result was obtained with the stratification by

schooling (up to incomplete secondary education, complete secondary education and higher education).

Discussion

Chronic diseases are associated with a worse QOL in a population. Several conditions can interfere in this perception including age, type of pathology, family support, beliefs, values and socioeconomic level^(14,15).

In relation to the characterization of the studied population, the analysis of the sociodemographic profile of sickle cell anemia patients showed a predominance of women and as sickle cell anemia is not a genetic disease that is influenced by gender, this finding may be explained due by the higher number of women in the population and by the fact that women seek information and health care services more frequently than men⁽¹⁵⁾.

Regarding ethnical data, most patients defined themselves as black; these results are in accordance with the other studies highlighting the close relationship between ethnicity and the disease⁽¹⁶⁾.

Most of the patients had incomplete high school, which is in accordance with the results obtained by Felix et al.⁽¹⁷⁾, in which 42.5% of sickle cell carriers had levels of schooling equal to or above complete secondary education. However, independent of the schooling, there was no improvement in the socioeconomic level, which is evident by the type of work, place of residence and the low remuneration of the participants in this study.

The marital status of the vast majority was single. According to the WHO, marital status of individuals influences the family dynamics and self-care. In one study, Pitaluga⁽¹⁸⁾ observed a positive correlation between loss of QOL from the psychological perspective and sickle cell anemia patients who lived alone.

It is evident that 68.8% of the patients were unemployed, showing the great impact of the disease in respect to this matter. This finding is in accordance with the literature. Pereira et al.⁽¹⁹⁾, in a survey of twenty-five sickle cell anemia carriers, observed that only 24% of them were employed and among those who were not, 72% indicated that the disease was the leading cause of the impediment.

In relation to the income, 87.5% of the interviewees had a per capita income of up to one and a half minimum wages coinciding with the study of de Silva et al.⁽²⁰⁾ in which 85% of the patients had a per capita income below this amount. It should be emphasized that that study dates back to 1993, and almost twenty years later, we observe the social and economic stagnation to which this population is submitted.

The results of this study show a low QOL and the involvement of the different domains analyzed. Grau Abalo⁽²¹⁾ shows that there is a natural instability in QOL construction as, in the course of a disease, patients can have different perceptions of their own QOL.

In the physical health component which includes functional ability, limitation by physical aspects, pain and general health status, the scores for limitation by physical aspects and functional ability, the most affected items, were below 30 with means of 26.56 ± 29.05 and 28.9 ± 16.69, respectively. This highlights the limitations in the daily activities and professional performance resulting from

the effect of the disease on physical health which causes a drop in the QOL. These findings are in accordance with Roberti et al.⁽²²⁾ whose study applied the WHO Quality of Life-Bref (WHOQOL-Bref) questionnaire to evaluate the QOL of 60 sickle cell anemia carriers. They found reduced scores for the physical domain due to dependency on medical treatment, the greater susceptibility to fatigue and pain, and reduced ability to work.

McClish et al.⁽²³⁾ applied the SF-36 to 308 sickle cell anemia patients and made a comparison with three cohort studies of other chronic diseases: asthma, cystic fibrosis and patients submitted to hemodialysis. They observed that sickle cell anemia carriers presented significantly lower scores than the general population in all domains except for mental health. In comparison with cystic fibrosis patients, sickle cell anemia patients also presented lower QOL for all domains except for mental health. Scores were similar for functional ability and mental health when compared to asthma patients, but they were worse for physical pain, vitality, social function and general health status. Regarding patients submitted to hemodialysis, the sickle cell anemia population had similar scores for physical limitation, emotional aspect, social function and mental health, lower scores for physical pain, general health and vitality and higher scores for functional ability.

Assis⁽²⁴⁾ performed a survey of 30 patients with sickle cell anemia, in which he evaluated QOL by applying the SF-36. He concluded that individuals did not present reduced levels of QOL.

With regard to physical health, the domain that presented one of the highest scores was general health status with a mean score of 56.09 ± 12.22 which supports the study of Roberti et al.⁽²²⁾ in which the patients defined the issues concerning general aspects of QOL and health as positive. The domain of pain showed a mean score of 50.31 and as pain crises are very common in these patients, it was not the domain with the lowest score.

The mental health item, which includes vitality, social and emotional aspects and mental health, showed that emotional aspects were the most affected with a mean score of 30.20 ± 33.18. This domain relates to problems with work or with other daily activities as consequences of emotional problems. Guimarães⁽²⁵⁾ claims that patients with chronic diseases are faced with changes in their lifestyles; their daily routines are changed. Thus, they need to adapt to a new lifestyle in which the continuous use of drugs, hospitalizations, and often, loss of working capacity are frequent.

Vitality was the domain with the highest score. This result shows that although sickle cell anemia patients have pain, fatigue and depressive symptomatology, they seek the best coping strategies against feelings of tiredness and exhaustion. According to Moreno-Jiménez⁽²⁶⁾, optimistic people develop more coping skills than those who are pessimistic. Health conditions get better when optimism is part of the individual's psychological condition.

The results of this study must be analyzed with caution due to the small sample and because it evaluated patients from a single health clinic.

Conclusion

Results obtained in this study show that the QOL is reduced in sickle cell anemia patients in all the different domains analyzed. The lowest scores were those related to functional ability, physical

limitations and limitations due to emotional aspects. The disease significantly interferes with the capacity of individuals to work so they generally have low-paid jobs.

It is necessary to perform further studies with samples from different locations, because knowing the sociodemographic profile and QOL of carriers is essential to improve the approach of health professionals and to assist public policy planning and implementation. This may improve the individuals' physical, psychological and socioeconomic realities. It is important to create strategies that allow better accessibility of sickle cell anemia patients to the job market and most of all to respect their physical and functional limitations.

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