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Physical and Mental Aspects of Patients With Sickle Cell Disease: a Portrait of Quality of Life in a Developing Country

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Abstract

OBJECTIVE This study aimed at analysing quality of life (QOL) indicators of patients with sickle cell disease (SCD) in treatment, investigating the epidemiological, socioeconomic and care situation provided to patients with SCD in the State of Maranhão, one of the poorest states of Brazil. METHODS: A cross-sectional study was carried out from March 2018 to February 2019, with the application of a generic quality of life questionnaire, SF-36, to patients attending a referral center for treatment of hemoglobinopathies in the State of Maranhão. 113 patients with SCD were interviewed and sociodemographic data, disease characteristics and laboratory tests (hemogram, foetal haemoglobin, DHL and reticulocytes) were collected. The SF-36 questionnaire was then applied. RESULTS: Most of the 113 patients were female, with a mean age of 26 years, declaring themselves to be of brown colour and living in the interior of the State. Most were unemployed, having low income and low schooling. About 92% were of the SS subtype, the most serious subtype. The percentage of neonatal diagnosis was only 27.4%. Regarding the SF-36 questionnaires, quality of life was classified as poor in relation to the physical component and good in the mental component. The use of hydroxyurea, the only medication approved in Brazil for the control and prevention of pain, promoted an improvement in the physical appearance of patients with SCD, howbeit, with no relation to the prevalence of clinical complications. CONCLUSIONS: The use of the SF-36 generic questionnaire showed impairment in the quality of life in the physical domains of patients with SCD, worsening in cases in which there was delayed diagnosis; in those individuals who claimed to have suffered prejudice; and in patients hospitalised for pain attacks. No deterioration of the mental components was observed. This scenario implies a need for government action sensitive to this public health problem.

Introduction

Since sickle cell disease (SCD) is a chronic and therefore debilitating disease, especially when it occurs in the most severe forms, it can interfere with patients' ability to perform their daily activities, causing temporary or permanent disability, as well as irreversible damage to the functioning of several organs, causing impairment to work and leading to decreased quality of life[1].

The World Health Organisation defines Quality of Life (QOL) as "the individual's perception of their position in life in the context of the culture and value system in which they live and in relation to their goals, expectations, standards and interests.[2] The term has been used by the population in general, but has stimulated countless studies conducted by professionals who provide assistance to people affected by chronic diseases, the prevalence and survival rates of which have increased in recent decades.[3,4]

Health-related Quality of Life has now become an important criterion in the evaluation of the effectiveness of interventions and treatments in the area. It has been used to evaluate the impact of chronic diseases on people's daily lives, and also to assess patients with the same disease but who present different criteria for the same treatment. In this evaluation, it is necessary not only to ascertain

the indicators of physical functioning, but also the social, mental and emotional aspects of the repercussions of symptoms and the individual perception of well-being.[5 - 8]

In order to evaluate QOL as an indicator of well-being, generic or specific instruments are usually used, which, through interviews conducted by professionals, generate values that serve as parameters for the identification and mapping of changes in various dimensions, such as the physical, psychological and social. General instruments related to QOL such as the SF–36 (*The Medical Outcomes Study 36-item Short Form Health Survey)* questionnaire for adults, and the PedsQL (*Pediatric Quality of Life Inventory)* for children, are used to measure physical, emotional, and social functioning and permit comparisons between individuals with SCD and healthy individuals. A specific PedsQL *Sickle Cell Disease* instrument, adapted for children diagnosed with SCD, has shown greater accuracy in evaluating QOL in the paediatric population.[9.10]

The SF–36 questionnaire (*The Medical Outcomes Study 36-item Short Form Health Survey)* was translated into Portuguese and validated by Ciconelli in 1999. This instrument was created with the purpose of being a generic questionnaire for health assessment, of easy administration and comprehension.[11] It is a multidimensional instrument consisting of 36 items, encompassed by eight components or domains: functional capacity, physical aspects, pain, general health, vitality, social aspects, emotional aspects, mental health, and a comparative evaluation between the current health conditions and that of one year before. It is of extreme importance in the knowledge of the patient's illness. This instrument measures both the negative (disease) and the positive (welfare) aspects.[12]

In this context, it is important to expand the discussions relating patient QOL to chronic diseases, such as SCD, an important public health problem in Brazil. The impacts on the patient's life are innumerable, coupled with various debilitating consequences. The understanding of these limitations allows us to identify problems and design appropriate interventions to modify variables that negatively interfere with the QOL [13.14].

Thus, the present study aimed at investigating and analysing QOL indicators of patients with SCD undergoing treatment. Considering all the characteristics of the SF–36 questionnaire and the growing interest in its application in clinical trials, it was decided to use this instrument in this study with sickle cell patients.

Methods

Study design and sample

This is a cross-sectional study carried out from March 2018 to February 2019, with patients with sickle cell disease seen at a referral center for treatment of benign hematologic diseases in Maranhão, one of the poorest states in Brazil, located in the Northeast region of the country. This research was authorised

and approved by the Research Ethics Committee of the Federal University of Maranhão—Brazil (Number 2.530.036).

Data collection occurred from March 2018 to February 2019, totalling 12 months. Patients' routine was not impaired by participation in the research, since this was done in the interval between the consultations.

Patients with 14 years of age or older, of both genders, were included. Diagnosis of Sickle Cell Disease was performed using the haemoglobin electrophoresis - High Efficiency Liquid Chromatography Technique, at any time, given physical, mental and intellectual conditions to communicate with the researcher, provided they agreed to participate in the survey.

Those under 14 years of age were excluded from the study, as well as patients with pain crises at the time of the interview; patients with psychiatric disorders, auditory or speech deficits; and those who did not consent to participate in the research.

After a detailed explanation of the study, the interviewee signed the Informed Consent Form (ICF). Participants under the age of 18 also signed the Term of Assent of Minors, and their guardians, the ICF.

By May 2017, there were approximately 700 patients diagnosed with SCD, of whom approximately 250 were older than or equal to 14 years of age, and therefore could be included in the present study. Thus, considering a sample of 111 patients with a 15% difference in quality of life scores between patients using hydroxyurea and non-users of this drug, we have a sample of 111 patients, with a sampling error of 5% and a confidence level of 95%. For this calculation, the following formula was used:

[Due to technical limitations, this equation is only available as a download in the supplemental files section.]

where:

n - calculated sample; N - population; Z - normal standardised variable associated with the confidence level; p - true probability of the event; e - sampling error.

Instruments for data collection

Sociodemographic and Clinical Characteristics Form

The volunteers were interviewed by the researcher, who filled out a sociodemographic form with information on: age, gender, marital status, skin colour, place of residence (urban or rural area), level of education, occupation, religion, monthly family income, and anthropometric data (weight and height).

Afterwards, information was gathered to characterise the disease, such as age at diagnosis, disease classification, disease prejudice, chronic complications (stroke, leg ulcers for more than 3 months,

priapism, myocardiopathy, osteomyelitis / aseptic necrosis (hypertension, diabetes mellitus, hepatitis B, hepatitis C, HIV), occurrence of painful crises, use of opioids, pulmonary hypertension, number of transfusions in the last 12 months, and information on the use of hydroxyurea.

Data from laboratory tests requested at a routine visit (haemoglobin concentration, leukocyte and platelet count, reticulocyte count, lactic dehydrogenase dosage, and fetal hemoglobin level) were recorded at the time of filling out the form.

Quality of Life Questionnaire - SF–36

To assess quality of life, the volunteers responded to the Brazilian version of the SF–36 questionnaire. The SF–36, translated into Portuguese and validated by Ciconelli in 1999, is a generic instrument of easy administration and understanding, which considers the individual's perception of their own health status and contemplates the most representative aspects of health. It consists of 36 items, subdivided into 8 domains: "functional capacity", containing 10 items - evaluates the presence and extent of limitations related to physical capacity; "physical aspects", 4 items - evaluates the limitations on the type and quantity of work, as well as the difficulties of doing work and activities of daily living; "pain", 2 items - evaluates the presence of pain, its intensity and its interference in activities of daily living; "general health status", 5 items - assesses how the patient feels in relation to their overall health; "vitality", 4 items - considers the level of energy and fatigue; "social aspects", 2 items - analyses the integration of the individual in social activities, "emotional aspects", 3 items - assesses the impact of psychological aspects on the well-being of the individual; "mental health", 5 items - includes questions about anxiety, depression, changes in behaviour or emotional loss of control; and psychological well-being. It also includes an item that evaluates health changes occurring in the period of one year, resulting in a worse or better health status.[11]

The data were analysed by transforming the responses of each domain into a score from zero (0) to one hundred (100).

Statistical analysis

The data were first stored using the Excel program. The results were presented in means, standard deviation, maximum and minimum values, and in frequency tables. To characterise the study group, descriptive analyses, comparisons and correlations between the variables were performed. The Kolmogorov-Smirnov test confirmed the normality of the data. The samples were compared using Student's t-test for two groups. For more than two groups, analysis was performed with univariate analysis of variance (One-way ANOVA). For statistical analysis, the GraphPad Prism 7 Program ® was used. The level of statistical significance was set at 5% ($p < 0.05$).

Results

In this study, 113 patients with sickle cell disease who were followed up were evaluated, in conformity with the criteria of inclusion. The clinical and sociodemographic characteristics are described in Table 1.

Regarding the characteristics of the disease, 92% of respondents had SCD classified as HbSS, and the remainder were divided between HbSC and HbS-β–0. Neonatal diagnosis of SCD was done by pricking the heel, in 27.43% of the sickle cell patients; 19.46% of the patients had diagnostic confirmation by age five; 27.43% between 6 and 15 years; and 25.55% only after 16 years of age.

The SF–36 quality of life questionnaires were first evaluated, all components of each domain being calculated and grouped into physical components (functional capacity, physical aspects, pain and general health) and mental components (vitality, social aspects, emotional aspects and mental health).

The means for each SF–36 sub-domain are shown in Table 2. Scores between 0 - 49 are classified as poor quality of life; in contrast, those between 50 - 100 are considered good quality of life. It can be observed that the worst scores were represented by the summary of the physical components (mean 48.19 21.51; among them, the physical aspect was the one with the lower averages.

There was no statistically significant difference between the genders, with p = 0.9273 for the summary of the physical components and p = 0.7597 for the summary of the mental components. However, when the sub-items are analysed in isolation, functional capacity and general health status were statistically significant, with p = 0.0056 and p = 0.0470, respectively (Table 3).

In relation to the age at which the diagnosis of SCD and QOL was made, it can be observed that there was a significant difference in the summary of the mental component, with a better QOL for those who had the earliest diagnosis of SCD, that is, at less than five years of age (p = 0.0066) (Table 4).

When questioned about having already suffered prejudice with regard to SCD, 32.74% confirmed that they had. For this comparison, there was a significant difference both in the summary of the physical and mental components, with worse quality of life for those who had already suffered this type of insult (Table 5).

Painful crises in the 12-month period were assessed. For that, groups of patients were formed according to the frequency of occurrence of pain in this period. No pain was reported by 12 patients; one to two crises in 41 individuals; and three or more seizures in 60 patient. The impact of pain can be verified by the inversely proportional relationship between QOL and the number of seizures, with a statistically significant p value for the summaries of both physical and the mental components (Table 6).

Of the 113 individuals who answered the questionnaire, 48, that is, 42.47% of the total, had been using hydroxyurea (HU) for more than 6 months. This drug is a ribonucleotide reductase inhibitor used to treat patients with SCD, because it increases the production of foetal haemoglobin, which protects against sickle cell crises. The average dose used was 24.16 mg / kg / day.

Patients were divided into groups according to the use of this medication for the treatment of SCD. It was observed that the SF–36 scores were better in the patients who used the medication, with statistical significance in the physical components (p = 0, 0101) (Figure 1).

The incidence of pain crises has been shown to decrease with the use of medication, but no statistically significant difference was found between groups (Table 7). It is noteworthy that opioid use to control painful seizures was observed in 56% of the patients.

Discussion

Chronic diseases can generate the development of physical, social and emotional problems, due to the organic dysfunctions of the underlying disease. SCD, in this scenario, also exhibits such characteristics. These complications can appear during their natural course, interfering directly in quality of life and leading to a decrease in life expectancy in this population.[15]

The sample investigated in the present study was comprised, in most of the cases, by women, by patients considered to be of a brown colour, of the SS genotype, having diagnosed SCD late (after five years) for the most part. Only 27.43% had been diagnosed at birth, well below expected. A study conducted in Cameroon, a country with worse socioeconomic conditions than Brazil, showed a median age at diagnosis of 4 years of life, also below that recommended by the CDC.[16] Belgium implemented the screening test for hemoglobinopathies in 1994, as well as one more database of national case registers in 2008, and already reaps fruits, with a mean age at diagnosis of 0.7 years, with an impact on the morbimortality of the disease.[17] It has been demonstrated that the early diagnosis of SCD, together with the capacity of a multiprofessional team and the participation of the family and the community, plays a central role in reducing complications, as well as prolonging the life span of carriers.[18]

In Brazil, in 2001, the National Neonatal Screening Program was instituted. The hemoglobinopathies test is performed as part of the neonatal screening test (foot prick test), based on Order No. 822/01 of the Brazilian Ministry of Health, for the early diagnosis of the pathology.[19] Thus, the results presented in this study reveal a certain fragility of the coverage of this screening test in our country, especially in the interior of the state, where 80% of the sample patients reside.

The mean low age for a chronic disease, 26 years, may reflect a reduced life expectancy for this group of people. It should be noted that the Human Development Index (HDI) for the state of Maranhão is the second lowest in Brazil (0.639)—which may also explain the lack of access to diagnostic tests and hospitals, which may influence the life expectancy of these patients. The median age of death for patients with SCD was 26.5 years in State of Bahia, 31.5 in State of Rio de Janeiro, and 30.0 in State of São Paulo. The current situation in Maranhão is comparable to that of Brazil in 1996, when the median age of death was 18.5 years.[20]

Considering that painful seizures begin as early as childhood and, as found in this study, are the most prevalent complications in this population, these young people learn to deal early with the disease and the limitations imposed by it. In the case of young people with SCD, hospitalisations limit their satisfactory school development, making them also stigmatised for being sick and absent.[21] In addition, SCD may create limitations on the practice of sports. For all these reasons, it is perceived that it is necessary to maintain the support of a multidisciplinary team with psychological and social support throughout the school life of this population. However, in this study, school delay can be inferred, and partly explained, by conditions of discrimination and lack of knowledge regarding the disease, and also by school absenteeism during hospitalisations for treatment of complications. Almost half of the patients did not reach the minimum of eight years of schooling, which is considered the minimum suggested by the Brazilian Federal Constitution.

The impact of clinical conditions on the professional life of the patients evaluated was evident in this study from the low income of the ill. This exposes them to a situation of social vulnerability. A large proportion of the population studied (69%) reported monthly incomes of up to two minimum wages. The occurrence of frequent hospital admissions to manage complications, such as painful crises and the presence of osteodegenerative alterations, creates problems for such patients to remain in paid work with a formal contract; this is evidence of the presence of multiple factors that make it difficult to perform activities that generate income. One study carried out in the United Kingdom, with a different socioeconomic situation, also showed the professional difficulties and lack of job stability generated by the limitations imposed by SCD.[22]

Another aspect that represents negative impact, leading to sadness and social isolation, is the prejudice and discrimination suffered by such patients. As verified in this study, patients with SCD were impaired, both in physical and mental components. This reality constitutes a deeply disturbing picture for most people with SCD, since this impairs their self-esteem, leading to the construction and perception of a negative self-image. [21]

The medians of the summaries of the physical and mental components on the SF–36 questionnaire among all interviewees (48.19 and 68.00, respectively) revealed that the physical impairment is more pronounced than the psychological. A similar study carried out in Cameroon in 2017 showed means of 47.3 and 41.0 for the physical and mental components, respectively, suggesting worse values in the latter, a finding different from that of the present research study; perhaps this could be explained by the unstable political and social situation in that country.[16] Another study, in Saudi Arabia, revealed even lower values than the Cameroonian study, probably because the SF–36 was even applied with patients during painful episodes.[23]

The domain of physical aspects, one of the items of the physical component of the SF–36, is composed of four questions (related to problems with work or some daily activity due to physical health).[11] Concerning question four of that domain, which refers to problems with a regular daily task as a consequence of physical health, it was observed that a large majority of the population had decreased

the amount of time spent on work, performing fewer tasks than they would like, and limiting their type of work or their chores, as well as referring to difficulties related to do their work or other occupations. Thus, this group of patients presented momentary incapacity for the performance of their activities, whether job-related or not, a fact that deserves attention—since they are of productive age for society.

Pain, and especially chronic pain, is now considered an important public health problem, with high prevalence in the adult population, being one of the main causes of temporary or permanent incapacity to work. Studies have shown that pain is the most persistent symptom reported in primary health care services, and is frequently accompanied by symptoms of depression and anxiety.[24] The results of this study show a negative correlation between quality of life of patients with SCD and pain (whether acute or chronic), i.e., the greater the intensity and symptoms, the worse is the quality of life of individuals. This was perceived in both the physical and mental domains. This deterioration is aggravated in proportion to the number of annual pain crises. Adequate control of this symptom was related to the lower rates of blood transfusions and lower frequency of hospital admissions in a year. It is important to note that a considerable variability in the frequency of painful crises between patients can be observed, depending on inherited sub-phenotypes. It has been demonstrated that haplotype GCH1, related to the nitric oxide metabolism, confers susceptibility to pain by altering endothelial function in patients with SCD.[25]

One aspect to be highlighted is that even in this high prevalence complication, the use of opioids, drugs indicated for the management of moderate or severe pain attacks by the WHO, occurs in just a little more than 50% of the cases. Several factors may explain this moderate use of opioids in patients with SCD.[26]

Opioids are drugs that can improve the quality of life of people with chronic pain, but which are mistakenly viewed with reservations by doctors and patients. Opiophobia is the fear of using or prescribing opioid drugs. In most cases, the phobia takes root through the myths about opium derivatives, associated with ignorance about their systemic effects. It has also been pointed out that the fear of using this type of analgesia may be related to a belief in the direct association of its prescription with the evolution of the disease or with the imminence of death.[27,28]

In addition, many health professionals fear that when prescribing such drugs, the patient will become chemically dependent. It is known, however, that when prescribing sub-doses of analgesics it predisposes patients to adopt a “drug seeker” behaviour, due to the fear of developing dependence. In this behaviour, the demand for the drug is driven by the fear of feeling pain and not having the medicine at their disposal.[27] Evaluation of pain is subjective, and requires the patient’s self-report. Often the health professional may tend not to rely on the description of the symptoms by the patient, associating such complaints with a possible psychological dependence. Despite the concern of health professionals, the percentage of psychological dependence in patients with sickle cell disease is only 1 to 3%.[26]

The appropriate management of episodes of pain attacks requires continual specialised training. It is important for the chronic patient pain approach to involve a multidisciplinary team to monitor the patient continuously. This prevents each professional from providing a different service, which would result in

uneven health care, without standardisation. Ensuring appropriate access to the drug improves the patient's relationship with the health care team and provides care based on the case history of each individual.[29]

Hydroxyurea (HU) is the only medication approved in Brazil for the prevention of pain crises, and its use is the best way to control complications.[30] It is known that HU has multiple effects on the erythrocytic lineage, promoting the elevation of HbF in 60% of the persons treated and increasing the haemoglobin level and the mean corpuscular volume, as well as reducing the number of reticulocytes. People who use it have half the seizures they had before the medication, and there is a decrease in the need for blood transfusions. Decreasing the frequency of seizures, patients require fewer hospitalisations, improving quality of life and lessening the overall cost of treatment.[31] It was observed in this study that only 42.47% of the patients used the drug, a percentage well below than expected, considering that more than 80% of the respondents had a history of pain crises. The explanation for this fact is that despite its availability through the Brazilian Unified Health System at no cost, the drug is not stored in the pharmacy of the referral blood bank; and it is up to the user to go to the State Pharmacy for Specialised Medicines in another neighbourhood, to apply, and receive a response after 15 days, forcing them to increase the amount of travel to the capital of the State, a fact that can delay or render unfeasible the onset of medication. Another stalemate would be the very resistance of prescribing physicians to prescribe medication and not being able to closely control the adverse effects thereof in patients residing in the interior of the state.

A Jamaican study involving 383 people with SCD with a mean age of 31 years andfollow-up at three years revealed that 66% were using HU with a mean dose of 21.5mg / kg / day. It has been demonstrated that organ damage and premature death still occur in spite of the use of HU, but that a suitable dose, preferably an increase to the maximum tolerated dose, is important, in order to achieve the desired effect[32]. It is noteworthy that this study used a mean dose lower than the present study.

One limitation of this cross-sectional study is that the chronic complications were reported by the patients, depending exclusively on the interviewee's memory—which may cause bias due to forgetfulness or even lack of knowledge about their complications. It is also worth noting that only one treatment centre was studied, with its own economic and social realities, and may be different from other economic situation, even within Brazil itself.

To date, there is no specific instrument to evaluate the quality of life of people with SCD among adults, but some generic instruments do serve this purpose. Further research is needed in order to increase the understanding of the impact of quality of life among adults with SCD. The quantification of QOL has a strategic function for the development and re-adaptation of clinical protocols, operational planning of health services, allocation of resources, and development, adaptation and strengthening of public health policies, especially in regions of high prevalence of the disease, such as the state of Maranhão - Brazil.

Declarations

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Authors’ contributions

CFAR, TAR and MSSC conceptualized the current study, conducted the analysis and drafted the manuscript. All authors reviewed and approved the final manuscript.

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Availability of data and materials

The datasets supporting the conclusions of this article are included within the article and its additional files.

Ethics approval and consent to participate

This research was authorised and approved by the Research Ethics Committee of the Federal University of Maranhão—Brazil (Number 2.530.036). The data do not contain any code that could identify respondents. All participants gave written consent for their participation.

Consent for publication

All participants gave written consent for the anonymised publication of data for research purposes.

Competing interests

All authors declare no competing interest.

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Tables

Table 1 – Sociodemographic characteristics of patients with sickle cell disease. São Luís. Maranhão - Brazil. 2019.

*Brazilian monthly wage in 2018: R$ 954,00; in 2019: R$ 998,00.

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		
Single	86	76.1
Married / Stable Union	26	23.1
Divorced	1	0.8
Skin Colour		
White	9	7.9
Brown	64	56.7
Yellow	4	3.5
Black	36	31.9
Residence		
São Luís - Capital	22	19.5
Countryside	91	80.5
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 x minimum wage	78	69.1
3 to 5 x minimum wage	11	9.8
> 5 x minimum wage	3	2.6

Table 2 - General quality of life of patients with SCD according to the SF-36 questionnaire. São Luís. Maranhão - Brazil. 2019 .

DOMAIN	Mean ± SD n= 113	Minimum value	Maximum value
Functional capacity	61.24 ± 22.93	10	100
Physical appearance	30.75 ± 42.65	0	100
Pain	64.98 ± 32.33	0	100
General state of health	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

Table 3 - Quality of life between genders of patients with SCD according to the SF-36 questionnaire. São Luís. Maranhão - Brazil. 2019.

DOMAIN	MEAN ± SD		p*
	Female n = 71	Male n = 42	
Functional capacity	56.69 ± 20.91	68.93 ± 24.36	0.0056
Physical appearance	32.04 ± 43.13	28.57 ± 42.26	0.6779
Pain	65.67 ± 31.61	64.35 ± 33.54	0.8338
General state of health	38.73 ± 19.45	30.83 ± 21.41	0.0470
Summary - Physical Components	47.82 ± 22.85	47.41 ± 22.76	0.9273
Vitality	53.45 ± 22.72	58.45 ± 22.59	0.2595
Social aspects	97.54 ± 74.78	85.71 ± 24.63	0.3236
Emotional Aspects	63.38 ± 45.13	54.76 ± 47.04	0.3363
Mental health	70.14 ± 22.95	75.33 ± 21.82	0.2391
Summary - Mental components	67.38 ± 24.19	68.85 ± 25.44	0.7597

* Student's t test

Table 4 - Quality of life and age at diagnosis of SCD using the SF-36 questionnaire. São Luís. Maranhão - Brazil. 2019

DOMAIN	MEAN ± SD		p*
	≤ 5 years n = 53	> 5 years n = 60	
Functional capacity	64.34 ± 27.17	58.50 ± 23.42	0.1778
Physical appearance	34.91 ± 44.49	27.08 ± 40.99	0.3328
Pain	83.82 ± 75.79	67.83 ± 32.10	0.0795
General state of health	41.51 ± 21.74	30.75 ± 17.99	0.0048
Summary - Physical Components	51.56 ± 23.43	44.32 ± 21.54	0.0899
Vitality	58.77 ± 20.07	52.25 ± 24.55	0.1280
Social aspects	90.33 ± 20.31	80.63 ± 22.72	0.0190
Emotional Aspects	80.51 ± 92.82	52.22 ± 46.89	0.0398
Mental health	76.91 ± 18.22	67.80 ± 25.21	0.0318
Summary - Mental components	74.54 ± 19.77	62.08 ± 26.95	0.0066

* Student's t test

Table 5 - Quality of life and prejudice against SCD according to the SF-36 questionnaire. São Luís. Maranhão - Brazil. 2019.

DOMAIN	MEAN ± SD		p*
	No n = 76	Yes n = 37	
Functional capacity	64.67 ± 22.28	54.19 ± 22.93	0.0219
Physical appearance	41.45 ± 45.38	8.784 ± 25.15	<0.001
Pain	75.86 ± 27.76	42.97 ± 29.56	<0.001
General state of health	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 6 - Quality of life and occurrence of pain crises in patients with SCD according to the SF-36 questionnaire. São Luís. Maranhão - Brazil. 2019

DOMAIN	MEAN ± SD			p*
	None n = 12	1 or 2 crises n = 41	≥ 3 crises n=60	
Functional capacity	80.42 ± 27.75	61.95 ± 20.40	56.92 ± 21.86	0.0043
Physical appearance	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 state of health	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 aspects	89.58 ± 12.87	89.33 ± 17.36	81.46 ± 25.08	0.1630
Emotional Aspects	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 7 - Painful crises at 12 months in patients with SCD. users or not of hydroxyurea. São Luís. Maranhão - Brazil. 2019.

Pain crises in 12 months	MEAN SD		p*
	With hydroxyurea n = 48	Hydroxyurea free n = 65	
No pain crises	8 (16.7%)	4 (6.2%)	0.8191
< 3 crises	18 (37.5%)	23 (35.3%)	0.5283
≥ 3 crises	22 (45.8%)	38 (58.5%)	0.2835

* Student's t test

Figures

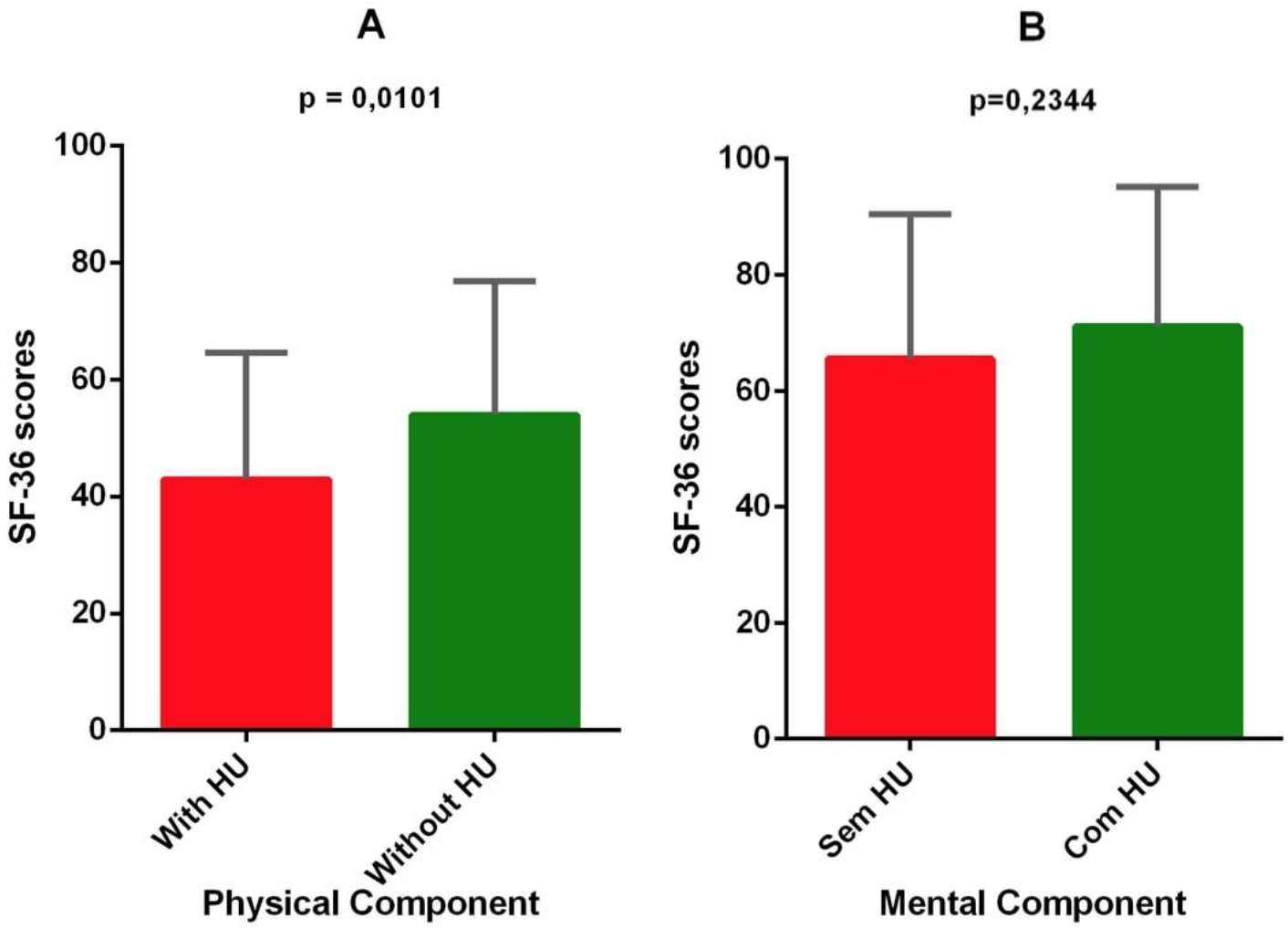


Figure 1

Scores on the SF-36 questionnaire and its distribution between the physical (A) and mental (B) domains in patients who did or did not use hydroxyurea. Results shown in mean ± SD. Value of p = 0.0101 between physical domains and p = 0.2344 between mental domains. considering patients using hydroxyurea or not. Student's t-test comparison.

Supplementary Files

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