ORIGINAL ARTICLE

Impact of hemophilia on health-related quality of life of patients accompanied by the blood center of Alagoas

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ABSTRACT

Quality of life is a critical aspect in the therapeutic plan of any patient, especially those who suffer from chronic diseases, such as hemophilia. Thus, this study aimed to assess the quality of life in patients with hemophilia treated by the Blood Center of Alagoas and describe the demographic and socioeconomic profile. 50 individuals were enrolled in the study, being over 18 years old and with hemophilia treated by the Association and the Blood Center of Alagoas. Data collection was carried out through interviews using two questionnaires: one on demographic and socioeconomic characteristics, and the other on health-related quality of life (Haem-A-QoL). Of this total, 100% were male, most aged between 20 and 40 years (80%), single (62%), living in the interior of Alagoas (58%), retired (56%), beneficiaries of retirement due to hemophilia complications (60%), with personal income in the last month of up to one minimum wage (78%) and without a private health plan (86%). The mean of the total Haem-A-QoL score was 40.08 (ranging from 0-96.46), with the worst performance in the domains "sports and leisure" (mean of 55.52) and "physical health" (mean of 50.16), and the best performance in the domain "partnership and sexuality" (mean of 17.48). In general, the hemophiliacs in the center studied reported good health-related quality of life. The Brazilian version of Haem-A-QoL proved to be a reliable instrument, with good internal consistency, revealing some problems faced by hemophiliacs that impact on HRQoL.

Keywords: Hemophilia, Health-related quality of life, Blood center.

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INTRODUCTION

Hemophilia is an inherited bleeding disorder caused by deficient activity of blood coagulation factors VIII (hemophilia A) or IX (hemophilia B). This coagulopathy is an X-linked recessive disorder, predominantly affecting men, who need only one affected X chromosome to manifest the disease[1].

Hemophiliacs have prolonged blood clotting time compared to a healthy individual, thus implying that these patients can bleed for a longer period, which can damage various tissues of the human body[2]. Severe hemophilia triggers joint or muscle bleeding associated with trauma, or without apparent cause. The moderate form causes bleeding usually related to trauma and procedures and, only occasionally, spontaneous. Mild hemophilia has as clinical manifestations bleeding associated with major trauma or procedures[3]. The main complication caused by the disorder is hemophilic arthropathy, a debilitating condition resulting from repeated joint bleeding that manifests itself as joint stiffness and chronic pain, leading to disability and compromising quality of life[4-5].

As a consequence, the health-related quality of life of hemophilia patients is affected by clinical manifestations of the disease that limit, depending on the severity, the patients' activities of daily living. Health-related quality of life (HRQoL) is a multidimensional concept on the impact of diseases on patients' lives. It can be influenced by how the individual deals with the illness, with treatment and with issues of access to care, considering the patient's perception of the impact of hemophilia on life. Indeed, although advances in science have enabled the cure or treatment of several disorders, medical intervention may cause pain, discomfort and wear to the patient, compromising the quality of life[6-7].

For this purpose, the Hemophilia Quality of Life Questionnaire (Haem-A-QoL), a disease-specific assessment tool, was used to investigate and determine the factors that interfere with HRQoL in this population, recognizing the most vulnerable to possible interventions of health services. Given the remarkable importance of quality of life in the medical context and the knowledge of complications caused by hemophilia, this study aimed to assess the health-related quality of life, identifying the axes that are impaired, and describing the demographic profile and socioeconomic status of patients with hemophilia treated at the Association of Hemophiliacs and the Blood Center of Alagoas (HEMOAL).

MATERIAL AND METHODS

This study has a prospective character and a quantitative approach. The project was submitted to the Research Ethics Committee of UNCISAL and was approved under protocol CAAE: 87587118.2.0000.5011. The research was developed at the Association of Hemophiliacs of Alagoas and at the Blood Center of Alagoas.

The sample consisted of patients over 18 years old with hemophilia who were admitted by the association during the period from September 2018 to April 2019 (except January 2019). All patients aged 18 years and over who had a confirmed diagnosis of hemophilia, regardless of type, and whose coagulopathy treatment was related to HEMOAL, were included in this research.

Data collection was carried out with individuals who agreed to participate in the study and signed the Free and Informed Consent Term (FICT). Posteriorly, the patients answered a questionnaire developed from two axes: one containing the demographic and socioeconomic characteristics, consisting of a modified version of the questionnaire developed by Ferreira (2012); and an assessment of health-related quality of life, elaborated through the Haem-A-Qol questionnaire. This questionnaire includes 46 items and 10 domains, namely: physical health, feelings, view of yourself, sports and leisure, work and school, dealing with hemophilia, treatment, future, family planning, partnership and sexuality.

The collected data were typed and organized into a Microsoft Excel 2010 spreadsheet, and processed in the Statistical Products and Service Solutions (SPSS) for Windows 12 program. For the demographic/socioeconomic data, simple descriptive statistics were used to calculate the absolute and relative frequencies of the variables. The score of each domain and the total score were transformed into values ranging from zero to 100, with zero representing the best HRQoL and higher scores representing greater impairment in HRQoL. Simple statistics were also calculated, describing the mean, maximum, minimum, standard deviation and Cronbach's alpha for each domain, highlighting those most affected by hemophilia. Importantly, Cronbach's alpha is a valuable tool used to investigate the reliability of the internal consistency of the research instrument. The results were displayed in table format.

RESULTS

Demographic and Socioeconomic Characteristics of hemophilia patients (Table 1)

The initial sample consisted of 52 patients. However, one individual was under 18 years old and other participant did not have a confirmed diagnosis of hemophilia, and were excluded from the study, based on the inclusion and exclusion criteria.

Of these 50 men with hemophilia, 40 (80%) were aged between 20 and 40 years, 6 (12%) were over 40 years old, and only 4 (8%) reported being younger than 20 years. Regarding marital status, the survey revealed that 31 (62%) patients were single, and 12 were married. 52% of respondents said they have no children.

Considering the household, 32 (64%) individuals reported living in municipalities in the interior of the state. Regarding accessibility to care facilities, 29 (58%) patients reported living more than 30 km from the Blood Center. The findings revealed a low level of education, which was reflected in the non-completion of primary school by 13 (26%) patients. When asked about occupation, 28 (56%) were retired, and 9 (18%) formal workers. As for income status, 39 participants (78%) declared personal income in the last month of up to one minimum wage.

Additionally, 30 patients (60%) declared themselves to be beneficiaries of retirement due to hemophilia, and 28 (56%) were unable to find a job after the benefit. Of the total, 43 patients (86%) reported not having a private health plan.

Table 1 - Characteristics of hemophilia patients at the Blood Center of Alagoas, 2018-2019

Variables	Classification	Absolute Frequency (N)	Relative Frequency (%)
Sex	Male	50	100
Age	20-40	40	80
	>40 years	06	12
	< 20 years	04	8
Marital Status	Single	31	62
	Married	12	24
	Stable union	07	14
Have children	No	26	52
	Yes	24	48
Residence	Interior of Alagoas	32	64
	Capital (Maceió)	17	34
Distance between home and blood center	More than 30 km	29	58
	Less than 30 km	21	42
Education	Incomplete Primary School	13	26
	Complete Primary School	13	26
	Complete High School	11	22
Occupation	Retired	28	56
	Formal Worker	09	18
	Student	05	10
	Informal worker	05	10
Personal income in the last month	Up to 1 minimum wage	39	78
	More than 1 minimum wage	06	12
Social security benefits for hemophilia complications	Yes	30	60
	No	20	40
Employee after receiving social security benefit	No	28	56
	Did not want to answer	18	36
Private health plan	No	43	86
	Yes	7	14

Source: Survey data, 2019

Table 2 - Means of scores, standard deviation and internal consistency of the Haem-A-QoL domains of hemophilia patients at the Blood Center of Alagoas, Brazil, 2018-2019

Domain	N	Mean	Minimum	Maximum	Standard Deviation	Cronbach's alpha
Sports and leisure	50	55.52	0	100	28.23	0.83
Physical health	50	50.16	0	96	31.93	0.94
View of yourself	50	44.40	0	100	23.43	0.49
Treatment	50	44.64	0	100	28.96	0.88
Future	50	23.20	0	95	27.43	0.87
Work and school	50	41.80	0	100	28.28	0.78
Feelings	50	43.30	0	100	31.95	0.91
Dealing with hemophilia	50	33.18	0	100	30.47	0.85
Family planning	50	47.14	0	87	19.90	0.84
Partnership and sexuality	50	17.48	0	86,66	24.53	0.85
Total	50	40.08	0	96,46	27.56	0.78

Source: Survey data, 2019

Table 3 - Mean scores and standard deviation of items in the domains with the greatest impact on the Quality of Life of hemophilia patients at the Blood Center of Alagoas, Brazil, 2018-2019.

Items in the "Physical Health" domain	Mean	Minimum	Maximum	Standard Deviation
I had pain in my joints	56	0	100	37.25
My swellings hurt	56	0	100	36.14
I had difficulty walking as far as I wanted to	54.40	0	100	38.55
It was painful for me to move	47.20	0	100	37.20
I needed more time to get ready	37.20	0	100	37.25

Items in the "Sports and Leisure" domain	Mean	Minimum	Maximum	Standard Deviation
I had to avoid sports because of my hemophilia	63.20	0	100	40.68
I had to plan everything in advance due to hemophilia	60.40	0	100	40.30
I was unable to play the sports I liked due to hemophilia	54.40	0	100	39.18
I was not free to travel where I wanted because of hemophilia	35.60	0	100	38.81

Source: Survey data, 2019

HEALTH-RELATED QUALITY OF LIFE ASSESSMENT

The mean Haem-A-QoL total score was 40.80, ranging from 0 to 96.46. The patients showed the worst performances in the domains "Sports and leisure" (mean of 55.52) and "Physical health" (mean of 50.16), reflected by the high scores obtained, indicating greater impairment in HRQoL. In the domain "Partnership and sexuality", the participants exhibited less impairment (mean of 17.48). Given that there is no standard score to classify the quality of life of patients in their respective domains, it is important to analyze that the values above the mean, that is, 40.8, are considered as items of impairment in quality of life, and domains with a mean below this value contribute favorably to the quality of life of the individuals under study.

The value of the minimum acceptable Cronbach's alpha coefficient to consider the questionnaire reliable is 0.7, below that the instrument is not considered consistent[8]. Usually, alpha values between 0.80 and 0.90 are preferred. The total alpha coefficient for the Haem-A-QoL reached 0.78, which points to an adequate reliability coefficient, suggesting the use of a good research instrument. In 9 of 10 Haem-A-QoL dimensions, the Cronbach's alpha ranged from 0.49 to 0.94 in the analysis of the internal consistency.

In the domain "View of yourself", the alpha was 0.49, showing that this item was not well correlated with the others in the questionnaire.

The domain "Sports and Leisure" reached the highest Haem-A-QoL mean. Among the items that had the worst scores, that is, with a mean closer to the maximum score, it is noteworthy the fact that the patients avoid sports (63.20), need constant planning due to hemophilia (60.40), and are unable to practice activities normally (54.40), impacting negatively on the quality of life. Regarding the items in the "Physical health" domain, the worst results can be observed for frequent joint pain (56) and swelling (56), accompanied by difficulty walking (54.40).

DISCUSSION

In the analysis of sociodemographic data, in relation to household, the majority of patients (64%) came from municipalities in the interior of Alagoas, evidencing the great demand for these institutions. Regarding accessibility to care facilities, 29 patients (58%) reported living more than 30 km from the Blood Center. This demonstrates that hemophilia patients deal with peculiar situations that affect their quality of life, as they face a geographic distance to carry out the treatment[9].

As for age, the most affected population is young adults between 20 and 40 years old (80%). There is a high percentage of retired patients, dependent on social security benefits due to complications of hemophilia (60%) and who were unable to find a job after the benefit (56%). These data are in accordance with a previous study conducted by Caio et al.[9], which reported a large number of hemophiliacs with an income of up to one minimum wage. Therefore, the average income of these patients is low, and it can be inferred that the disorder can contribute to the maintenance of this situation.

Only 12% of patients were older than 40 years old. This is possibly due to the lower life expectancy of hemophiliacs until the end of the last century and the high mortality decades ago. Until then, treatment was absent or inadequate, with the risk of transmitting hepatitis virus and the acquired immunodeficiency syndrome (AIDS) due to contaminated blood components. In fact, there was no strict control of blood components and derivatives, and the viral inactivation technique was developed only in the 1980s[4,10-11].

Regarding marital status, 31 (62%) patients reported being single. This result is similar to the findings achieved by Garbin et al. (2007), who associated this fact with the difficulties of acceptance of hemophiliacs and the physical limitations resulting from clinical manifestations[12].

Moreover, 43 patients (86%) do not have private health plans, hence demonstrating the key role of a quality public health system in maintaining treatment for this population group.

In our study, the mean HRQoL score was 40.08 among adult hemophilia patients. Notably, Ferreira et al.[13] indicated a mean HRQoL score of 35.33 for the individuals under study, similar to the result observed in research with Greek patients, with a mean of 33.59[14], indicating better QoL scores in Haem-A-QoL. However, patients with hemophilia in Iran had worse scores when compared to patients in Alagoas, Minas Gerais and Greece, with a mean score of 51.07[15].

Among the few studies published discussing HRQoL in adults with hemophilia, one in Brazil used the Haem-A-QoL as an assessment tool. The cross-sectional analysis was conducted in Minas Gerais by Ferreira et al.[13], and evaluated 39 adults with hemophilia who presented relatively higher scores in the domains "Sports and leisure" (mean of 49.89) and "Physical Health" (mean of 43.30), indicating worse HRQoL in these dimensions. In the domain "Partnership and sexuality" (mean of 17.52), the participants had a better quality of life, similar to that described in this study.

Remarkably, the domain "Sports and Leisure" reached the highest average, reflecting the high prevalence of arthropathy found in these patients. The routine of pain, deprivation and uncertainties contribute to the worsening quality of life. Joint pain and swelling make it difficult for these patients to walk and move, and they can develop musculoskeletal complications[16]. Furthermore, the individuals avoid sports and deprive themselves of travel for fear of not being able to control possible bleeding.

Nevertheless, physical exercise and physical therapy are essential to reduce pain, correct motor deficits, increase muscle strength, improve mobility, functionality and joint stability. Thus, engaging in these activities helps to improve the quality of life of hemophiliac patients and is a trend that should be encouraged[17-18].

The domain "Physical health" revealed the second highest mean, being also greatly influenced by the presence of arthropathy, which worsened the HRQoL of hemophiliacs. This result can be correlated with the high percentage of patients dependent on social security benefits due to complications of the life-threatening condition.

Ferreira et al.[13] found in prior research a better Haem-A-Qol score in the domains "Dealing with hemophilia" and "View of yourself", suggesting a possible better quality of life in their group of individuals in comparison to the patients evaluated in this study, in which the domains "Partnership and sexuality" and "Future" showed lower means, indicating less loss of HRQoL in these dimensions.

Moreover, the individuals exhibited greater impairment of "physical health" when compared to "sexuality". This could be because the physical impairment does not interfere, in the same proportion, with the sexuality of these patients. In turn, another possibility is the omission of answers in this domain or the fact that the questionnaire excludes part of the sample with regard to sexual preference, since it uses the following statements: "I have had difficulty dating women because of hemophilia" and "I have felt insecure in my relationships with women because of hemophilia"[19]. This bias was not corrected at the time of application of the questionnaire.

The dimension "Treatment" showed the worst mean (44.64) in comparison to the study carried out in Minas Gerais (35.47). Therefore, it is worth emphasizing the importance of specialized and qualified services and professionals, regardless of whether they are from the Blood Centers or not, so that they know how to deal with the complications of hemophilia, as trust in doctors and safe treatment improve HRQoL.

CONCLUSION

In general, patients with hemophilia showed good HRQoL in the study. The Brazilian version of the Haem-A-QoL, answered by the individuals, proved to be a reliable instrument with good internal consistency. This demonstrated that numerous aspects are influenced by the disorder, thereby interfering with the quality of life, such as low income, difficulty and distance to access treatment services, pain and few job opportunities after diagnosis. Thus, when applied, the questionnaire helps in the development of individualized care, based on the patients' perception of the impact of hemophilia on their life, not taking into account only the strategies that the health professional considers to be effective.

The dimensions "Sports and Leisure" and "Physical Health" showed to significantly affect the quality of life, influenced by arthropathy, the major complication of the disease. Therefore, physical activity, physical therapy and other rehabilitation and muscle strengthening modalities are pivotal to reduce the symptoms of hemolytic arthropathy and improve the HRQoL of these patients.

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