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Original Article

Socio-demographic, economic and health profile of adults with sickle-cell disease

Perfil sociodemográfico, econômico e de saúde de adultos com doença falciforme

Perfil sociodemográfico, económico y de salud de adultos con enfermedad falciforme

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Objective: to describe socio-demographic and economic characteristics, lifestyle, clinical manifestations, use of medications and monitoring of adults with sickle-cell disease. **Methods:** a descriptive study with quantitative approach, made with 20 adults, registered in a Hematology and Hemotherapy Center, using, for data collection, interviews in the participant's home and medical records. In order to have the database, the EpiDatae analysis software through statistical program was used. **Results:** most of the population consisted of women, married, with complete high school, which used exclusively the Unified Health System. The average age was 30.6 years, and 90.0% (95% CI 68.3-98.8) were black. All of them reported painful crises and fatigue. They used folic acid daily 35.0% (95% CI 15.4-59.2). **Conclusion:** the implications of sickle-cell disease could be mitigated through primary, secondary and tertiary health care, according to the needs of those adults.
Descriptors: Anemia, Sickle Cell; Adult Health ; Epidemiology.

Objetivo: descrever características sociodemográficas e econômicas, hábitos de vida, manifestações clínicas, medicamentos em uso e acompanhamento de adultos com doença falciforme. **Métodos:** estudo descritivo com abordagem quantitativa, realizado com 20 adultos, cadastrados em um hemonúcleo, utilizando-se, para coleta de dados, entrevista no domicílio do participante e prontuários. Para a confecção do banco de dados, foi utilizado o *software* EpiData e análise por programa estatístico. **Resultados:** a maioria da população foi formada de mulheres, casadas, com Ensino Médio completo, que utilizava exclusivamente o Sistema Único de Saúde. A média de idade foi de 30,6 anos, e 90,0% (IC95% 68,3-98,8) eram negros. Todos relataram crises álgicas e fadiga. Estavam em uso diário de ácido fólico 35,0% (IC95% 15,4-59,2). **Conclusão:** as implicações da doença falciforme poderiam ser minimizadas por meio de cuidados de saúde condizentes às necessidades desses adultos em serviços de atenção primária, secundária e terciária.
Descritores: Anemia Falciforme; Saúde do Adulto; Epidemiologia.

Objetivo: describir características sociodemográficas y económicas, estilo de vida, manifestaciones clínicas, uso de medicamentos y acompañamiento de adultos con enfermedad falciforme. **Métodos**: estudio descriptivo, con abordaje cuantitativo, con 20 adultos, registrados en un hemonúcleo, utilizándose para recolección de datos entrevista en casa y registros del participante. Para confección del banco de datos, se ha utilizado el *software* EpiData y programa de análisis estadístico. **Resultados**: mayoría de la población compuesta por mujeres, casadas, con enseñanza secundaria completa, que utilizaban exclusivamente el sistema de salud. Edad media de 30,6 años, y 90,0% (IC del 95%: 68,3. -98,8) negros. Relataron crisis dolorosas y fatiga. Estaban en uso diario de ácido fólico 35,0% (IC del 95%: 15,4-59,2). **Conclusión:** las implicaciones de la enfermedad falciforme podrían minimizarse mediante atención de salud según las necesidades de estos adultos en servicios de atención primaria, secundaria y terciaria
Descriptores: Anemia de Células Falciformes; Salud del Adulto; Epidemiología.

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Introduction

The term "sickle-cell disease" comprises a set of hereditary hemolytic anemia that has, in general, the presentation of hemoglobin s inside the red cell. It is one of the most common hereditary diseases in the world. In Brazil, due to the large presence of afrodescendant population, the sickle-cell disease is a group of relevant diseases and disorders[1].

Concerning the incidence of sickle-cell disease, Minas Gerais is the third state in Brazil in this raking, followed by the states of Bahia and Rio de Janeiro[1]. It is estimated that about 3,500 children with sickle-cell disease and 200,000 with sickle-cell trait are born in the country per year[2].

In adults, the main signs and symptoms are chronic anemia, painful crises, infections, cerebral vascular accident, ocular complications, gallstones, priapism, ulcers in the lower limbs, impaired growth, delay of secondary sexual characteristics, menarche and delayed first ejaculation[2].

The treatment of sickle-cell anemia, as a whole, presents different hierarchical complexities. It is based on hydration, fight against pain, oxygen therapy, fighting infections, folic acid intake (because it is an important vitamin in the formation of red blood cells), blood transfusion, and in many cases, the use of hydroxyurea medicine, which is associated to the increase of intravascular production and nitric oxide infra-erythrocytes, which makes vasodilatation easier. When eligible, the bone marrow transplantation can also be made[1,3].

Because it is a chronic disease, the treatment is made throughout life and to be successful, the families, from diagnosis, should learn about the signs of complications and how to act properly in different complications[4-5], for individuals who do not have adequate social, economic and family support tend to have more problems resulting from the disease and from the difficulties of adjusting to it.

Thus, sickle-cell disease has hematologic, clinical, genetic, anthropological and epidemiological importance, among others, due to its morbidity and its high mortality rate[6-7].

Because of all that, the importance of widening the knowledge of health, socioeconomic and demographic aspects, involving a person with sickle-cell disease is perceived, because the impacts it brings to the patient's life are numerous, together with several limitations. Understanding these limitations allows identifying problems and devising appropriate actions of intervention to modify variables that negatively affect the well-being and quality of life of those people[4]. Such knowledge can subsidize the nursing professional for more specific educational activities to promote health and prevention of diseases of the person with sickle-cell disease.

Considering the severity, frequency and the factors that influence the evolution of sickle-cell disease, this study aimed at describing the socio-demographic and economic characteristics, lifestyles, clinical manifestations, medications in use and the assistance to adults with sickle-cell disease.

Method

This study was a part of a research project entitled "Completeness of care for patients with sickle-cell disease: home care and the strengthening of the net of support of a Hematology and Hemotherapy Center located in the state of Minas Gerais." This is a descriptive study with quantitative approach.

The study setting was the home of people with sickle-cell disease, older than 18 years of age, registered in the regional center of Hemominas, of the Foundation Center of Hematology and Hemoterapy, in the state of Minas Gerais, located in Divinópolis, MG and residents in this county. Hemominas is the foundation responsible for the health related to hematology and hemotherapy in Minas Gerais, with regional units

(blood banks), regional centers (Hematology and Hemotherapy Center) and contracting hemotherapy units. Data collection was carried out between August 2013 and January 2014.

Based on convenience sampling, the following selection criteria were adopted: adults with sickle-cell disease, registered and assisted in the Hematology and Hemotherapy Center. Exclusion criteria were people with sickle-cell disease under 18 years of age, without address and/or telephones in the register, unable to understand and answer the questions due to intellectual disability, and being assisted in another blood center. In the register of that Hematology and Hemotherapy Center, 25 adults were identified with sickle-cell disease who met the inclusion criteria.

At first, the researchers sent a correspondence inviting all adults selected to participate in the research and, after 15 days, telephone contact was made to explain the research objectives and to have the consent of the participant. Two people selected refused to participate in the research, a subject was in prison and two of the addresses provided were not found; therefore, 20 adults were part of the study. The interviews were held at the residence of the patients, having been previously scheduled through phone calls, at the times they made available by them.

Data were obtained through a questionnaire prepared by the authors and previously tested, consisting of open and closed questions. The variables studied included demographic and socioeconomic characteristics of the patients (sex, age, type of sickle-cell disease, housing conditions, level of education, current occupation, marital status, number of household members, family income, religion, number of rooms, type of residence, and whether the houses had running water and septic tank).

In order to have a databank, the EpiData software, version 3.1b was used, and for the data analysis, the Statistical Package for the Social Sciences, version 18.0 was used. A descriptive analysis was made and the respective Confidence Intervals (95% CI) were calculated for the point estimates of this sample.

The project was approved by the Committee of Ethics of the Universidade Federal de São João del Rei – Campus Centro-Oeste, having as co-participant the Fundação Hemominas, according to legal opinion number 599.680-0.

Results

20 adults with sickle-cell disease were interviewed, registered at the Hematology and Hemotherapy Center researched, being 55.0% (n = 11; 95% CI 31.5-76.9) were female. The age ranged from 20 to 61 years, with a predominance of women between 30 and 39 years (50.0%; 95% CI 27.2- 72.8), the average age was 30.6 years and 90.0 % (n = 18; 95% CI 68.3 - 98.8) were black.

It was observed that 5.0% (n = 1, 95% CI 0.13 - 24.9) of sickle-cell disease patients had no education, while 65.0% (n = 13; 95% CI 40.8- 84.6) had high school degree and 20.0% college degree (n = 4, 95% CI 5.73 - 43.7). Most of them were married (50.0%; 95% CI 27.2 - 72.8) and 20.0% (95% CI 5.73 - 43.7) of the participants were unemployed at the time of the study. It was also observed that most adults with sickle-cell disease were in classes C1 (45.0%; 95% CI 23.1 - 68.5) and C2 (35.0%; 95% CI 15.4 – 59.2), while 5.0% (n = 1, 95% CI 0.13 - 24.9) were in class D. These data are shown in Table 1.

Table 1 - Socio-demographic and economic characteristics of adults with sickle-cell disease. n=20

Characteristics	n (%)	CI (95%)*
Age (years)		
20-29	8 (40.0)	19.2 – 63.9
30-39	10 (50.0)	27.2 – 72.8
≥40	2 (10.0)	1.24 – 31.7
Color of the skin		
Black	18 (90.0)	68.3 – 98.8
Other	2 (10.0)	1.24 – 31.7
Marital status		
Married	10 (50.0)	27.2 – 72.8
Single	7 (35.0)	15.4 – 59.2
Stable union	3 (15.0)	3.21 – 37.9
Schooling		
Illiterate	1 (5.0)	0.13 – 24.9
Grade School	2 (10.0)	1.24 – 31.7
High School	13 (65.0)	40.8 – 84.6
University	4 (20.0)	5.73 – 43.7
Occupation		
Employed	11 (55.0)	31.5 – 76.9
Unemployed	4 (20.0)	5.73 – 43.7
Housewife	4 (20.0)	5.73 – 43.7
Student	1 (5.0)	0.13 – 24.9
Economic Classification**		
B1	1 (5.0)	0.13 – 24.9
B2	2 (10.0)	1.24 – 31.7
C1	9 (45.0)	23.1 – 68.5
C2	7 (35.0)	15.4 – 59.2
D	1 (5.0)	0.13 – 24.9

*Exact Binomial Interval;**Critereon of Economic Classification in Brazil,2014[8]

All of the participants had access to water supply and basic sanitation facilities. As for religion, 15 (75.0%; 95% CI 50.9 - 91.3) people were catholic, 5 (25.0%; 95% CI 8.66 - 49.1) were evangelical. Concerning family arrangements, 10 (50.0%; 95% CI 27.2 - 72.8) of the participants lived only with their spouse or partner, 6 (30.0%; 95% CI 11.9 - 54.3) lived with the parents and brothers, 3 (15.0%; 95% CI 3.21 - 37.9) lived with spouse and children and 1 (5.0%; 95% CI 0.13 - 24.9) adult with sickle-cell disease lived with his grandmother.

Considering the characteristics of the underlying disease, it was found that the average age at diagnosis of sickle-cell disease was 5.2 years, and 5 (25.0%; 95% CI 8.66 - 49.1) participants had their diagnosis after 7 years of age.

Regarding the type of the most common hemoglobinopathy found, the homozygous of the hemoglobin s/HbSS was present in 18 (90.0%; 95% CI 68.3 - 98.8) adults and HbSC in 2 (10.0%; 95% CI 1.24 - 31.7). During the course of the disease, 5 (25.0%; 95% CI 8.66 - 49.1) people reported to have already had a transfusion. Regarding lifestyle, 1 (5.0%; 95% CI 0.13 - 24.9) adult reported being a smoker and 2 (10.0%; 95% CI 1.24 - 31.7) reported the use of alcoholic beverages. As for physical activity, 2 (10.0%; 95% CI 1.24 - 31.7) of the adults with sickle-cell disease reported exercising daily.

When being inquired about medications in use, 7 (35.0%; 95% CI 15.4 - 59.2) participants reported daily use of folic acid and 3 (15.0%; 95% CI 3.21 – 37.9) reported not taking any medicine (not even in painful crises), 1 (5.0%; 95% CI 0.13 - 24.9) participant had hypertension and used folic acid as oral amlodipina hypotensive, 8 (40.0%; 95% CI 19.2 - 63.9) adults with sickle-cell disease reported to use complementary therapies in painful crises, 5 (25.0%; 95% CI 8.66 - 49.1) used heat as a complementary form of treatment, 2 (10.0%; 95% CI 1.24 - 31.7) massage and 1 (5.0%; 95% CI 0.13 - 24.9) associated phytotherapy to painkillers (ibuprofen and acetaminophen) in painful crises.

Regarding the use of health services for monitoring of sickle-cell disease, more than half of the participants (55.0%; 95% CI 31.5 - 76.9) reported exclusively the services available through the Unified Health System in the three levels of care. These data are described in Table 2.

Table 2 - Types of hemoglobinopathy, lifestyle, treatment and type of health services used by adults with sickle- cell disease. n = 20

Variables	n(%)	CI(95%)*
Types of hemoglobinopathy		
HbSS	18 (90.0)	68.3 – 98.8
HbSC	2(10.0)	1.24 – 31.7
Lifestyle		
Smoking	1 (5.0)	0.13 – 24.9
Alcoholism	2 (10.0)	1.24 – 31.7
Physical exercises	2 (10.0)	1.24 – 31.7
Blood transfusion		
Yes	2 (10.0)	1.24 – 31.7
No	18 (90.0)	68.3 – 98.8
Medications in use		
Folic acid	7 (35.0)	15.4 – 59.2
Folic acid + hydroxyurea	1 (5.0)	0.13 – 24.9
Folic acid + opioid + non-steroidal analgesics	2 (10.0)	1.24 – 31.7
Folic acid + non-steroidal analgesics	3 (15.0)	3.21 – 37.9
Folic acid + hypotensive	1 (5.0)	0.13 – 24.9
Folic acid + opioid	1 (5.0)	0.13 – 24.9
Hydroxyurea + non-steroidal analgesics	1 (5.0)	0.13 – 24.9
Non-steroidal analgesics	1 (5.0)	0.13 – 24.9
None	3 (15.0)	3.21 – 37.9
Complementary therapy		
Massage	5 (25.0)	8.66 – 49.1
Heat	2 (10.0)	1.24 – 31.7
Phytoteraphy	1 (5.0)	0.13 – 24.9
None	12 (60.0)	36.1 – 80.9
Use of health services		
Unified Health System (primary, secondary and tertiary health care)	11 (55.0)	31.5 – 76.9
Unified Health System + secondary health care (Hematology and Hemotherapy Center)	9 (45.0)	23.1 – 68.5

* Exact Binomial Interval

All the participants reported having painful crises; 15 (75.0%; 95% CI 50.9 - 91.3) showed incapacitating pain to carry out daily activities. Regarding the frequency of these painful crises, 7 (35.0%; 95% CI 15.4 - 59.2) participants reported they occurred monthly; 5 (25.0%; 95% CI 8.66 - 49.1) reported a biannual basis; 2 (10.0%; 95% CI 1.24 - 31.7) weekly; 1 (5.0%; 95% CI 0.13 - 24.9) participant reported painful crises daily; 1 (5.0%; 95% CI 0.13 - 24.9) person reported crises quarterly and another (5.0%; 95% CI 0.13 to 24.9) annually. Adults with sickle-cell disease who were not using drugs daily (15.0%; 95% CI 3.21 - 37.9) reported moderate level of pain.

The support to carry out their daily activities during painful crises was reported by 12 (60.0%; 95% CI 36.1- 80.9) participants; 8 (40.0%; 95% CI 19.2 - 63.9) reported they did not have any support during these crises. All participants reported fatigue throughout the year, and 15 (75.0%; 95% CI 50.9 - 91.3) participants reported the intensity of fatigue as moderate, 5 (25.0%; 95% CI 8.66 - 49.1) participants reported that fatigue was monthly; 4 (20.0%; 95% CI 5.73 - 43.7) daily; 3 (15.0 %%; 95% CI 3.21 - 37.9), weekly; 3 (15.0%; 95% CI 3.21 - 37.9), quarterly; and 3 (15.0%; 95% CI 3.21 to 37.9), annually. 2 (10.0%; 95% CI 1.24 - 31.7) reported fatigue each semester.

Regarding priapism, 2 (10.0%; 95% CI 1.24 - 31.7) men reported episodes along their lives. As for delayed menarche, 6 (30.0%; 95% CI 11.9 - 54.3) women reported having menstruation after 16 years of age; 5 (25.0%, 95% CI 8.66 -49.1) adults with sickle-cell disease reported a delay in the beginning of the development of secondary sexual characteristics, such as appearance of pubic hair, development of the breast in girls and low pitch voice in the boys (Table 3).

Table 3 - Clinical manifestations presented by adults with sickle-cell disease. n = 20

Clinical manifestations	n(%)	CI(95%)*
Priapism	2 (10.0)	1.24 – 31.7
Premature ejaculation	4 (20.0)	5.73 – 43.7
Delayed menarche	6 (30.0)	11.9 – 54.3
Delay in the secondary characteristics	5 (25.0)	8.66 – 49.1
Painful crises	20 (100.0)	-
Level of pain during the crises according to the visual analogical scale of pain		
Light (0-2) – does not interfere in DA**	-	
Moderate (3-7) – interferes, but does not incapacitate DA	5 (25.0)	8.66 – 49.1
Intense (8-10) – incapacitates DA	15 (75.0)	50.9 – 91.3
Fatigue	20 (100.0)	-
Level of fatigue		
Light (0-2) – does not interfere in DA	2 (10.0)	1.24 – 31.7
Moderate (3-7) – interferes, but does not incapacitate DA	15 (75.0)	50.9 – 91.3
Intense (8-10) – incapacitates DA	3 (15.0)	3.21 – 37.9

* Exact Binomial Interval **DA = Daily Activities

All participants, at some point in life, suffered splenic sequestration crisis, there were 8 (40%; 95% CI 19.2 - 63.9) participants with dactylitis and other 8 (40%; 95% CI 19.2 - 63, 9) participants reported tonsillitis at some point of life. Ulcers of the lower limbs and jaundice were mentioned by 8 (40%; 95% CI 19.2 - 63.9) participants. 3 (15.0%; 95% CI 3.21 - 37.9) adults with sickle-cell disease were affected by urinary tract infection, 6 (30.0%; 95% CI 11.9 - 54.3) reported infections of the respiratory tract at some point in life. Gallstones was reported by 7 (30.0%; 95% CI 11.9 - 54.3) participants, all of which required surgical intervention (cholecystectomy). 1 reported ocular complications during their lifetime and 1 adult with sickle cell disease reported osteomyelitis.

Discussion

The lifestyle and health conditions of a population characterize the way a person is placed within the social world. Such characteristics are confirmed by socioeconomic, political and cultural factors that affect the environment, behavior and biology of these people, acting in their health/disease condition and therefore in their well-being and their quality of life. These social determinants provide significant influence in the lives of people with chronic disease, such as the patient with sickle-cell disease, for presenting great vulnerability. Facing this situation, these people require care and monitoring by health services in order to protect them and provide them with better quality of life[9-11].

In addition to the chronicity of the disease, there is a close analogy of sickle-cell disease with the black ethnic group, with double stigmatizing character, resulting from the presence of the disease and the ethnic origin. However, there is still association between diagnosis and prevalence among afrodescendant people. Therefore, these people are socially marginalized and have significant loss of quality of life[9, 11-12]. The assumption of the close relationship between the black ethnic group and sickle-cell disease was evidenced in this study and is consistent with the literature[6-7].

In the analysis of socio-demographic and economic data, it was observed that most of the participants, were female and most of them were within the age range 30-39 years. Only one person was older than 60 years, emphasizing the average age of patients with sickle-cell disease, which is 48 years in the country[1]. The life expectancy of people with sickle-cell disease is reduced to around 25-30 years compared to the general population without sickle-cell disease, due to the complications of sickle-cell disease such as kidney failure, vaso-occlusive crises, acute chest syndrome and cerebral vascular accident[13]. According to a study made with 264 patients with sickle-cell anemia with severe pain crisis, during an average period of monitoring of 4.92 years, 40 patients died[11]. The high lethality, mainly among youngsters, demonstrates the severity of the disease and its great epidemiological importance, reflected in the low life expectancy of these people.

Because it is a genetic disease not related to

sex, there are few publications addressing gender in sickle-cell disease. In this study, there was a majority of women, due to the small sample size. In a survey made in Goiás, similar data were found, that is, 53.3% of participants were female[10].

In this study, it can be seen that most of the participants had homozygote of the hemoglobin s/HbSS, which is also consistent with the literature, which shows that this is the most common, yet being the most severe genotype, usually associated to low life expectancy[1,12]. Half of the study participants were married and lived only with their spouse. This datum was not observed in other studies. In a survey made in Brazil with adults with sickle-cell anemia, 71.7% of the participants were single and lived with their parents [1].

It was noticed that there was a predominance of high school education in the participants and the monthly income was equals to or under four minimum wages, a little more than half of the participants reported to be employed. In a study also made in Minas Gerais it was found that the majority (59.1%) had high school level and 37,0% had registered jobs, besides the low-income (74.1% under three minimum wages) [14]. As a consequence of these socio-economic data, most of the participants (55.0%) used exclusively the Unified Health System.

It is known that early diagnosis promotes taking preventive measures, which may interfere positively in the treatment and in the course of the disease. This study highlighted that the average age of the diagnosis was 5.2 years, a similar age observed in another study[3]. It is important to highlight that all study participants were 20 years old or older and, only after 2001, early diagnosis of sickle-cell disease and other hemoglobinopathies in newborn infants is made trough neonatal screening. That exam covers almost of the newborns (98.0%) in Minas Gerais[15].

The clinical manifestations of sickle-cell disease are quite variable and resulting from the sickled cells, which strongly influence the microcirculation blood flow, making them adhere to the wall of the blood vessel. The consequences of the adherence are characterized by blood vessel occlusion and reduced blood flow in the capillaries, resulting in venous stasis and hypoxemia that lead to acute pain crises and progressive chronic organ tissue injury[3].

All study participants reported painful crises and, in relation to fatigue, most reported being moderate, but with monthly or lower frequency, confirming other studies[3,9]. The painful condition is closely related to secondary tissue ischemia to the sickling of red blood cells. Other factors that may contribute are occurrence of vaso-occlusive crises, activation of endothelial cells, adhesion of erythrocytes and leukocytes, vasoconstriction, coagulation activation, cellular dehydration, inflammatory response and damage to the blood flow.

Therefore, acute episodes of pain or vaso-occlusive crises are a trademark of sickle-cell disease with economic impact due to the high cost of health care. Severe painful vaso-occlusive crises continue to be a marker for the severity of sickle-cell disease and premature mortality along other known risk factors for death, including high-speed tricuspid regurgitation, high ferritin and impaired renal function[11].

The data obtained in this study confirm that the sickle-cell disease, once it is chronic and has a course with wide clinical variability, causes limitations in the life of the patient, being pain, fatigue and other symptoms most likely responsible for the physical and emotional destabilization of the person.

The triggering mechanism of sickle-cell disease pain is always complex and probably heterogeneous, according to the place of occurrence. Thus, the pain of dactylitis corresponds to an inflammatory process initiated by necrosis of the bone marrow in the distal portions of the limbs and the chest syndrome involves vaso-occlusion and infection. A study made in the United States reported that 35.0% of people with sickle cell disease older than 20 years have been hospitalized due to recurrent pneumonia[10], confirming the data from this research.

Two men, from a total of nine patients, had

priapism. The literature shows that priapism occurs in approximately 30.0% of men. Adults with sickle-cell disease and priapism history have five times higher risk of developing pulmonary hypertension[14].

Studies showed that 50.0% of patients with sickle-cell disease have delayed sexual development[10,13,15]. Such datum was not evident in this study, which can be attributed to the small sample size.

The incidence of lower limb ulcers was 40.0%, but the literature shows that this can vary between 25.0% and 75.0%[16]. Another important aspect is the variation of incidence in different age ranges; this complication appears only after the second decade of life[1].

All the participants who reported having had gallstones needed surgical intervention (cholecystectomy), which differs from the literature, probably because the sample was too small. In a survey made in São Paulo, 60.0% of the people who underwent cholecystectomy had association with hemopathy, especially sickle-cell anemia and spherocytic anemia [17].

Only one study participant made use of hydroxyurea, which was introduced as a treatment for sickle-cell anemia/HbSS for more that 25 years, based on its ability to increase the level of fetal hemoglobin . Hydroxyurea is the biggest inhibitor of polymerization of deoxy-Hb and thereby prevents the sickling of red blood cell, chronic hemolytic anemia, vaso-occlusive painful crises, heart attack and necrosis in various organs, improving clinical and life expectancy of people with sickle-cell disease[18].

There is a consensus that hydroxyurea should be more widely used. A study made in the United States, which monitored people who had been using that medicine for more than 17 years, showed that the drug is safe and that its use also appears to be related to reduced mortality. There was also reduction of the incidence of painful crises and hospitalizations, and the probability of survival in 10 years was 86.0% in treated cases compared to 65.0% among untreated[18].

Complementary therapies on painful crises were reported by 8 participants. Of these, most adults with sickle-cell disease used thermotherapy (in the case, the use of heat) to help relieve the pain. The use of heat in relieving pain promotes muscle relaxation and a sensation of comfort; vasodilation and increased blood flow occur, which favors local immunological contribution of defense cells and cytokines that accelerate the resolution of the anti-inflammatory process - in the case of sickle-cell disease, there is improvement of ischemia[19]. Currently, there are complementary therapies, that can mean improvement in the assistance to the users of the Unified Health System, as they provide another form of treatment and prevention of diseases[20]. Acupuncture and music therapy, for example, are complementary therapies that can be used for the benefit of people with sickle-cell disease.

Data in the literature on alcoholic beverage consumption and tobacco use in people with sickle-cell disease were not found. However, it is known that these vices could cause additional damage to their health, such as cancer and liver cirrhosis. In this study, only one person smoked for 30 years and two participants reported drinking alcoholic beverages.

The survey also showed that more than half of the adults with sickle-cell disease only used the services of the Unified Health System. It is important to highlight that, after the diagnosis of sickle-cell disease, patients are systematically monitored by the nearest Hematology and Hemotherapy Center[16]. In the age range of the study participants, the assistance at the Hematology and Hemotherapy Center was provided every six months[16]. It is highlighted that, due to the chronic nature of sickle-cell disease, it is essential to have shared assistance with the services of primary health care through an effective system of reference and counter reference, with the establishment of a care plan in order to promote the link with both the services and the integrality of care.

This study had as limitation the reduced sample size because it was made with adults with

sickle cell disease treated in only one average size county of Minas Gerais, and also the register of some patients was also outdated at the Hematology and Hemotherapy Center researched. However, this limitation serves as a guide for more detailed future studies and should foster this type of evaluation with larger groups of patients in different counties and states of our country, for a possible confirmation of these preliminary results.

Conclusion

In adults, the sickle-cell disease can go through a wide clinical variability and provoke limitations in people's lives, such as; pain, fatigue and the need to search for health services which are probably responsible for the decrease in quality of life and well-being.

The disease reflected negatively on daily activities and on the quality of life of the adults with sickle-cell disease. However, difficulties in coping with the disease, low quality housing, unemployment and / or poorly paid job could be minimized with social and psychotherapeutic treatment, which should be made available to the patients.

The person with sickle-cell disease should get more attention from the health care and social promotion, which will certainly reflect on improvement of the quality and life expectancy of the same. The results of this study can be used by nurses to support the planning of work in order to promote health of adults with sickle-cell disease.

Collaborations

Amaral JL contributed in the collection, organization, analysis and interpretation of the data, writing of the article and final version to be published. Almeida NA and Santos PS contributed in the collection, organization, analysis and interpretation of the data. Oliveira PP and Lanza FM assisted in the elaboration of the article and helped in the revisions.

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