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Clinical conditions of patients with hemophilia assisted in a regional hemocenter: a cross-section study

ABSTRACT | The aim of the study was to characterize the sociodemographic and clinical profile of patients with hemophilia followed at a Regional Blood Center. Epidemiological cross-sectional study with data collection by document analysis with 30 records from a Regional Blood Center in the Center-South region of Ceará. Statistical analysis was performed using Microsoft Office Excel® 2016 software. The work was approved by the Research Ethics Committee. There was a predominance of males (27), aged 20 to 59 years, single marital status, incomplete elementary education, residing in rural areas and income below 01 minimum wage. As for the clinical profile, hemophilia A in severe form, has a sedentary lifestyle, with a history of transfusion procedures. Most have two or more associated clinical manifestations. With the prevalence pointed out among the participants, the need for comprehensive and equitable assistance is evident, with an emphasis on nursing actions.
Keywords: Hemophilia; Bleeding; Epidemiology.

RESUMEN | El objetivo del estudio fue caracterizar el perfil sociodemográfico y clínico de los pacientes con hemofilia seguidos en un Centro Regional de Sangre. Estudio epidemiológico de diseño transversal con recolección de datos por análisis documental con 30 registros de un Centro Regional de Sangre de la región Centro-Sur de Ceará. El análisis estadístico se realizó con el software Microsoft Office Excel® 2016. El trabajo fue aprobado por el Comité de Ética en Investigación. Predominó el sexo masculino (27), de 20 a 59 años, estado civil soltero, educación primaria incompleta, residente en área rural e ingresos por debajo de 01 salario mínimo. En cuanto al perfil clínico, la hemofilia A en forma severa, tiene un estilo de vida sedentario, con antecedentes de procedimientos transfusionales. La mayoría tiene dos o más manifestaciones clínicas asociadas. Con la prevalencia señalada entre los participantes, se evidencia la necesidad de una atención integral y equitativa, con énfasis en las acciones de enfermería.
Palavras claves: Hemofilia; Sangrado; Epidemiología.

RESUMO | O objetivo do estudo foi caracterizar o perfil sociodemográfico e clínico de pacientes com hemofilia acompanhados em um Hemocentro Regional. Estudo epidemiológico de delineamento transversal com coleta de dados por análise documental com 30 prontuários de um Hemocentro Regional da região Centro-Sul do Ceará. Realizou-se análise estatística através do software Microsoft Office Excel® 2016. O trabalho foi aprovado pelo Comitê de Ética de Pesquisa. Predominou o sexo masculino (27), com idade de 20 a 59 anos, estado civil solteiro, ensino fundamental incompleto, residência em zona rural e renda inferior a 01 salário mínimo. Quanto ao perfil clínico, a hemofilia A na forma grave, apresentam sedentarismo, com histórico de procedimentos transfusionais. A maioria com duas ou mais manifestações clínicas associadas. Com a prevalência apontada entre os participantes, evidencia-se a necessidade de uma assistência integral e equânime, com ênfase nas ações de enfermagem.
Palavras-chaves: Hemofilia; Hemorragia; Epidemiologia.

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INTRODUCTION

Hemophilia is an inherited pathology of blood clotting, resulting from the defect of clotting factors (CF), causing inability to contain bleeding. It is considered a chronic disease with physical and psychological consequences, even disabling, depending on the severity of the injury and the level of CF deficiency. [1]

The most common symptoms of hemophilia are hemorrhages. These can be classified as mild, moderate and severe and occur mainly in the joints, muscles, mouth and nose. They are related to mortality when they affect the Central Nervous System, the gastrointestinal system, the throat or when they are secondary to severe trauma. [2]

The worldwide geographical disposition of hemophilia corresponds to approximately one case for every 7.500 people with type A (HA) hemophilia, and one case for every 35 thousand people with type B hemophilia (HB), both of which are aimed at births of male individuals . In Brazil, HA is estimated in about 80% of cases, being more frequent than HB. According to the 2016 Hereditary Coagulopathies Profile, Ceará had 4,82% (488) cases of AH and 2,56% (51) cases of HB. [3]

It constitutes the most prevalent type of coagulopathy, representing an important public health problem, because in addition to demanding a high cost due to treatment and prophylaxis, it also affects the hemophiliac globally. The disease compromises not only its physical aspects, but also social and psychological ones due to its long-term complications, affecting the family and other subjects involved in their daily lives. Such a condition can be better assisted by knowing the profile of the patients to better target health care strategies. [4]

Studies that characterize the sociodemographic profile of people with hemophilia are necessary because they aim to get to know them in their illness process. In such a way it is possible to establish a comprehensive care plan with the appropriate guidelines and appropriate treatments, easing the burden that the disease exerts on the hemophiliac and his family. [7]

Thus, the objective of the study is to characterize the sociodemographic and clinical profile of patients with hemophilia followed in a blood center with regional coverage.

METHOD

This is an epidemiological study with a cross-sectional design, based on primary databases. The study was carried out in the coagulopathies department of the Regional Blood Center of Iguatu, located in the Center South Region of the State of Ceará. The choice of the research field is justified by the fact that the aforementioned institution is a reference in the service of Hematology and Hemotherapy in the region, accounting for the blood policy and for the medical care of hematological diseases in 28 municipalities, covering the 17th and 18th Health Regions. and, therefore, providing assistance to a population of approximately 643.686 inhabitants, which suggests an expansion of the research field. [5]

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The regional blood center has 30 records of registered patients, thus configuring the study population. The research sample is of the census type, that is, it consists of the 30 medical records of patients diagnosed with hemophilia. The diagnosis was confirmed through specific laboratory tests for this genetic condition. The sampling took place through the application of the inclusion criteria: medical records of hemophiliacs registered in the Regional Blood Center of Iguatu-CE and medical records of patients undergoing clinical follow-up, having performed at least one average consultation at the Regional Blood Center of Iguatu-CE. The medical records of patients who had other coagulopathies and patients who died were excluded from the sample.

Epidemiological information was collected using document analysis technique, using the medical records of hemophiliac patients, obtaining the following data: sociodemographic and clinical.

Data collection occurred by searching the dossiers of hemophiliac patients registered in November 2016, the dossiers have different names in the Blood Center. The study population groups documents with information on sociodemographic data and clinical screening. For the collection,

TABLE 1- Sociodemographic profile of hemophiliac patients attended at the Blood Center of Iguatu-CE in 2016

Variável	N	%
Sexo		
Feminino	3	10
Masculino	27	90
Idade		
02–19	12	40
20–59	14	47
60–77	4	13
Estado civil		
Solteiro	16	53,3
Casado	14	46,6
Escoralidade		
Analfabeto	9	30
Infantil	1	3
Fundamental Completo	2	7
Fundamental Incompleto	13	43
Médio Completo	2	7
Médio Incompleto	3	10
Moradia		
Zona urbana	10	33
Zona rural	20	67
Renda familiar		
Menos de um salário	14	47
Salário mínimo	13	43
Mais de um salário	3	10
Total	30	100%

Source: Iguatu-CE, 2020. HEMOCE / IGUATU-CE, 2016

TABLE 2- Clinical and functional conditions of patients attended at the Regional Blood Center of Iguatu-CE in 2016, Iguatu-CE, 2020.

Variável	N	%
Tipo de hemofilia		
Tipo A	22	73
Tipo B	8	27
Gravidade da hemofilia		
Leve	7	23
Moderada	7	23
Grave	16	53
Faz parte do Programa de Dose Domiciliar		
Sim	27	90
Não	3	10

a form was used with dichotomous and multiple choice questions regarding the sociodemographic and clinical profile of each patient.

The variables included in the study were: both sexes, marital status (all), age group (2-77 years), residents in urban and rural areas, education up to high school, all types of hemophilia severity and who have already received transfusion blood.

In the analysis phase, descriptive statistics were used using Excel 2016. The data were presented through tables with absolute and relative frequencies.

The procedures and conduct adopted in the present study followed the ethical and legal recommendations of Resolution No. 466, of December 12th, 2012. The study was approved by the Ethics and Research Committee of the Universidade Regional do Cariri - URCA, obtaining the opinion number: 1.827.856 and CAAE: 58595116.0.0000.5055.

RESULTS

Considering the number of registered medical records of patients with hemophilia in the studied locus, there was predominance: men 27 (90%), adults aged 20 to 59 years (47%), single (54.3%), who attended elementary school incomplete (43%), live in rural areas (67%) and survive on a family income below the minimum wage (47%).

Table 2 shows the clinical and functional characteristics of the patients. Hemophiliacs treated at the blood center of Iguatu-CE have hemophilia A (73%), being of severe magnitude (53%), receiving a home dose (90%), having been transfused (70%), presenting two or more clinical manifestations (83%), has osteoarticular complications (67%) and is sedentary (83%).

Já foi submetido a transfusões clínicas		
Sim	21	70
Não	9	30
Ocorrêcia das manifestações clínicas		
Nenhuma	3	10
1 manisfestação	2	7
2 ou mais manifestações	25	83
Presença de Complicações Osteoarticulares		
Sim	20	67
Não	10	33
Prática de Atividade Física		
Sim	5	17
Não	25	83

Source: HEMOCE / IGUATU, 2016.

DISCUSSION

The majority of hemophiliac patients in the study were male 27 (90%). This finding is explained by the pathophysiology of the disease itself, showing that hemophilia is transmitted from mother to child, as a genetic inheritance. [6,7] The prevalent age group in the study ranged from 20-59 years (47%), corroborating with studies that show that the population with this age group tends to be the most affected by the disease. [7,9]

The research pointed out that the majority of hemophiliacs were single, it can be inferred that this finding stems from the young age of the patients, so that many end up spending more time with their parents through dependence on their care. [8]

A low level of education was revealed, given that they receive the diagnosis early and even before they start attending school. Due to the severe nature of the disease, hemophiliacs tend to isolate themselves from social life because they are limited to practicing vigorous activities and playing. However, the disease does not cause any intellectual disability, making it indispensable that not only the health team, but also the family, stimulate them to live with other individuals, especially in the school environment. [9]

Regarding the geographic origin of the patients followed, most are located in rural areas, causing difficulty in the process of getting to the city for consultations and treatment. A study carried out in Pelotas found that 41,7% lived in the municipality where the regional blood center was located, the remaining 58,3% lived in other municipalities. [7]

Data on income were out of date according to the minimum wage that is currently provided, R$ 1.045,00. 10 Records notify that there was the receipt of benefits for those with physical impairment, but did not contain the amount received. HA occurred more frequently in the study, being in agreement with other studies that show that this corresponds to 80-85% of cases in Brazil. [3,7,11]

In the profile outlined by this study, the severity of hemophilia was shown to be severe, corroborating with studies carried out in other locations, such as in the Regional Blood Center of Pelotas-HEMOPEL. [7] In a survey conducted in Colombia, it was found that in a sample of 33 hemophiliac patients, the severity of the disease at the severe level represented 82%. [12]

The main and safest method currently used for treatment is the administration of FC VIII and IX, being suitable for types A and B respectively. These work as a way to supply the lack of CFs not produced by the hemophiliac organism. [13-14] Since 1999, the Ministry of Health through the Home Dose Program aims to make the hemophiliac more autonomous and participative in terms of their therapy. It also provides training to patients or caregivers for the infusion of CF at home through venoclysis. [13-14] The study showed that 90% of hemophiliacs participate in the program.

Another method also used as a form of treatment is the transfusion of blood products. The risks involved consist of adverse effects or transfusion reactions, which may occur during or after the transfusion. Serious transfusion reactions are more rare, with mild ones being the most common and present during the nursing practice routine. [15,16,17]

The analyzed medical records brought as clinical manifestations: hemarthrosis, hematuria, melena, hematemesis, menorrhagia, bleeding after trauma and dental procedures, postpartum bleeding, oral and skin bleeding and epistaxis. In severe stages, episodes of spontaneous hemorrhage predominantly in muscles and joints. However, they do not usually bleed, and there are cases of patients who never report bleeding problems. [18]

Regarding osteoarticular complications, these include atrophies and proliferation of bones and irregularities of the articular surface with osteophyte formation, accelerated development and overgrowth of the epiphysis caused by excessive blood flow, bone necrosis and cystic formation, as well as impediment of bone growth resulting from the interference of the bone nutrition. The joints

that are frequently affected are those of the ankle, knee, hips, shoulder, elbow and wrist. These complications affect the joints that are used in the performance of basic activities, at the risk of irreversible sequelae in the osteoarticular apparatus, loss of limb movement. [19,20]

Activities that involve running and jumping can trigger several osteoarticular complications, which can lead to serious sequelae. Thus, it remains for the hemophiliac to adhere to permanent treatment and obtain an improvement in their quality of life. The research patients are sedentary, reinforcing the problem of hemophiliacs regarding the compromise of constant support and movement structures, such as the joints. [20-21]

Faced with this problem, it is important to offer guidance on the regular and adequate practice of exercises, as these will be beneficial for the proper functioning of the osteolocomotor system, preventing the patient from becoming sedentary and consequently having future complications. [20-21]

CONCLUSION

Hemophiliacs need comprehensive and differentiated assistance due to their particularities. For this, it is necessary to produce studies that seek to know the profile of the hemophiliac and his disease, to subsidize the therapy by the professionals. Among these, the nurse stands out with his ability to plan health care actions taking into account the individuality of the subject and involving the family, the team and the other social segments in the knowledge about the disease and the ways of dealing with it.

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