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SOCIODEMOGRAPHIC CHARACTERISTICS OF MEN WITH HEMOPHILIA IN THE SOUTH OF BRAZIL[1]

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ABSTRACT

The objective was to describe the sociodemographic characteristics of men with hemophilia in monitoring by a Regional Blood Center located in southern Rio Grande do Sul, Brazil. It is a qualitative and descriptive study, whose data collected between June 2014 and November 2015. Intensive interview technique applied with field notes and data analysis was descriptive. Participated 12 men with severe hemophilia and aged between 19 and 55 years, most with hemophilia A, are married, living in municipalities in the south of the Rio Grande do Sul, with infectious diseases, joint sequels and history of family history of hemophilia who evolved to death, and half of them children, and disability retirees.

Keywords: Hemophilia A. Hemophilia B. Chronic disease. Male. Nursing.

INTRODUCTION

Hemophilia A is a hereditary hemorrhagic disease linked to chromosome X in which the person is deficient or abnormal clotting activity of factor VIII or IX of blood coagulation, called hemophilia A or hemophilia B, respectively. In about 70% of cases, it transmitted only to the male carrier mother, but in about 30%, manifestation does not occur in other family members, which gives the new mutation of the phenomenon. It is estimated the prevalence of hemophilia A in a case every 5,000 to 10,000 births male and hemophilia B in a case every 30,000 to 40,000 male births[1].

In 2016 were 184,726 people with hemophilia worldwide. Countries with populations between 185 and 324 million people, among them Brazil, Nigeria, Pakistan, Indonesia and the United States had, in that order, 12,119, 308, 1,343, 1,954 and 16,949 people with hemophilia[2]. According to the Ministry of Health, in 2015 Brazil and the State of Rio Grande do Sul had, respectively, 11.856 cases and 674 people with hemophilia, with 9,908 and 587 with hemophilia A and 1948 and 87 with hemophilia B[3].

It recognized that hemophilia is a chronic disease with no cure, and intravenous infusion of factor VIII or IX is essential for users who have disabled one of these factors or abnormality in their coagulant activity. These factors may be derived or recombinant human plasma, which produced by molecular biology techniques and highly purified[1].

Furthermore, hemophilia generates physical complications such as muscle and joint bleeds, which are the most frequent. However, other less recurrent bleeding can be extremely serious and even fatal, as in the central nervous system. Swedish study that evaluated the quality of life of users with hemophilia A and B concluded that they have a lower quality of life and have the most impaired mental health than physical health[4]. Given that, these bleeding events cause high morbidity and have implications for the quality of life of users with hemophilia[5].

Thus, it highlights the costs involved in treating the disease. An Italian study indicated that the average annual cost per user was 117,732.00 euros in 2012[6]. In the United States, a study found that the treatment of hemophilia cost 17 277 US dollars per person per year and it is estimated that the total amount spent on health plans in the treatment of American users with hemophilia may reach half a million dollars[7].

Studies characterizing users with hemophilia point to the importance of knowing them and their illness, looking for a full service, information, proper guidance and treatments, helping to reduce the burden of disease on them and their family[8-9]. As well as the costs of health systems[7].

Still, hemophilia is a disease that manifests itself in

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men and women with and most often asymptomatic, not suffering with the symptoms and treatment. Given the above, this study is justified by believing in the importance of describe the sociodemographic characteristics of men with hemophilia, in order to assist health professionals to know them, understand their actions and contribute to the quality of treatment and their lives. As well as the social and family environment, to assist in the understanding of the disease and care. In this sense, the objective was to describe the sociodemographic characteristics of men with hemophilia in monitoring in a Regional Blood Center located in southern Rio Grande do Sul, Brazil.

METHODOLOGY

This is a qualitative descriptive study, because it aims to study the characteristics of a particular group: distribution by age, sex, origin, education level, income level, physical and mental health status, among others(10). The 12 male participants met the inclusion criteria: being male, have hemophilia A or B, have aged 18 years and be user accompanied by a multidisciplinary team in the Pelotas Regional Blood Center (HEMOPEL); and the exclusion criteria: to present cognitive limitations, such as speech, attention and / or reasoning, and do not speak the Portuguese language. The initial approach to men was made in person or by phone and meetings for interviews previously agreed, the place indicated by them, therefore occurred in HEMOPEL or in their homes.

The HEMOPEL is a referral service for people with bleeding disorders residents in the coverage area of the 3rd and 7th Regional Health Coordination (CRS) of Rio Grande does Sul State. Users receive outpatient multidisciplinary care by medical, nursing staff, physiotherapy, and dentistry. Monitoring involves consultations, tests, infusion, and distribution of blood clotting factors.

Data collection took place from June 2014 to November 2015, through intensive interviews and field notes that contributed to the corpus of data analysis and the construction of this characterization, and data analysis was descriptive(10).

The study complied with the ethical principles for research involving human beings, according to the National Council of Health n. 466/12(11). The Research Ethics Committee of the State Production and Research Foundation approved its development for Health, under Opinion No. 671,719 on 3 June 2014. The Consent and Informed (IC) signed in duplicate, being one with the participant and another to the researcher. IC were presented in the study objectives, the guarantee of anonymity, free access to data and results and the freedom to withdraw from participation at any time and without any damage. Participants identified by the letter "H", the hemophiliac man, followed by Arabic numerals corresponding to the chronological order of interviews, plus the age.

RESULTS AND DISCUSSION

The study included 12 men with hemophilia. It found that the participants were aged 19-55 years old, with a mean age of 38.6 years. Most (9 - 75%) had a history of family history with hemophilia who progressed to death (only family H8 not progressed to death). In most hemophilia A and all have serious form of the disease, there was a predominance of infectious diseases, especially hepatitis C (7 - 58.3%), and sequelae joint (10 - 83.3%) (Table 1).

Regarding the level of schooling, five (41.7%) had incomplete primary education and the other high school degree (2 to 16.7%) or university (2 to 16.7%) and ongoing (3-25%). Regarding the profession / occupation, half (6-50%) received retirement because of illness. A total of seven (58.3%) participants resided in other municipalities other than the city of Pelotas is located where the reference center for treatment, and were married, and six (50%) had one to four children (Table 2).

Hemophilia A occurs more frequently than hemophilia B and represents around 80% of cases(1). Other studies[2,8-9,12-14] also show the predominance of hemophilia A. Both classified according to the activity level of clotting factors VIII and IX. It considered severe when the plasma levels of these factors is below 1%, moderate between 1 and 5% and lightweight 5 to 40%[1]. In the most common serious form of bleeding is hemarthrosis and hematoma muscle bruising and trauma related to, or commonly, spontaneous[1,8]. The gravity, other studies corroborate the results found in this work, with a higher occurrence of severe form[8,12,14].

Regarding age, prophylactic treatment significantly improves the quality of life of users and minimizes bleeding risky life[15-16], with a positive impact on life expectancy. In recent years, there has been a considerable increase in life expectancy of users, and that caused the emergence of morbidities arising with age, such as cardiovascular and kidney disease, diabetes mellitus and osteoporosis[1]. Another study also showed morbidities such as heart disease, depression, hypertension, and hepatitis C, among others[13].

As for infectious diseases, mostly hepatitis C was

present in young and in the elderly. This complication, often because of contaminated blood products and / or blood products, according to studies[12-14]. In the 1980s and early 1990s, Products without viral inactivation, blood components and first-generation blood products, caused high mortality rate due to transmission to users with hemophilia the hepatitis B virus (HBV) and C (HCV), and human immunodeficiency virus (HIV)[1,17]. It is important to note that over the years have been developed and adopted tracking conditions to prevent the transmission of diseases.

PARTICIPANT	AGE	A FAMILY HISTORY OF HAEMOPHILIA	MORBIDITIES	SEQUELS
H1	24 years	Brother and extended family	Severe hemophilia A and hepatitis C.	Chronic synovitis in the lower limbs
H2	49 years	Brother and extended family	Severe haemophilia A, hepatitis C and hypertension	Hemophilic arthropathy in upper and lower limbs
H3	55 years	Brothers and grandnephew	Severe haemophilia A, hepatitis C and heart problem	Hemophilic arthropathy in upper and lower limbs
H4	28 years	Brother and extended family	Severe haemophilia A	Not
H5	52 years	Brothers and grandson	Severe haemophilia A	Hemophilic arthropathy in upper and lower limbs
H6	51 years	Nephew and great-nephew	Severe Hemophilia B, Hepatitis C, Chagas disease, hypertension and diabetes	Hemophilic arthropathy in upper and lower limbs
H7	19 years old	Has family history	Severe haemophilia A	Synovitis in chronic lower limb
H8	45 years	Nephew	Severe haemophilia A	Hemophilic arthropathy in upper and lower limbs
H9	22 years	Has family history	Severe haemophilia A	Not
H10	39 years old	Uncle	Severe haemophilia A, C hepatitis, cholelithiasis, hypertension, anxiety and depression	Hemophilic arthropathy in upper and lower limbs and sequel after muscle hematoma
H11	39 years old	uncles	Severe hemophilia A and hepatitis C	Hemophilic arthropathy in upper and lower limbs
H12	40 years	Uncle, nephews and cousin	Severe Hemophilia B and hepatitis C	Hemophilic arthropathy in the lower limbs and severe muscle atrophy secondary to arthropathy

Table 1. Characteristics of participants according to age and family and clinical variables. Pelotas, Brazil, 2015 (n = 12).

The results in this study showed that most respondents had sequelae, such as hemophilic arthropathy in the lower limbs, most commonly in knees and ankles, corroborating the results of another study, which showed high prevalence of arthropathy, influencing the quality of life of users[8,13]. Still, with respect to the presence of joint involvement, researchers found that 43% of users did not have to articulate change, 25% had 3 or more joints and 16% in one and two joints[9].

The hemarthrosis affect most often the joints of the knee, ankle, elbow, shoulder and hip, and when recur in the same joint can cause joint degeneration, referred to as hemophilic arthropathy. This result in a progressive and irreversible inflammation of synovial tissue (synovitis), as well as degenerative cartilage lesions due to the presence of intra-articular blood. Its main characteristics are the loss of motion, the fixed flexion contracture and muscle atrophy secondary to disuse as well as pain[1].

The data obtained in this study showed that 5 (41.7%) men resided in the municipality of Pelotas and the rest (58.3%) in other municipalities of the 3rd and 7th CRS of the Rio Grande do Sul State, Brazil, served by HEMOPEL. Studies have found that most users with hemophilia also resided in other regions, not to the municipality where the reference center for the treatment[8-9]. The fact that users move to other municipalities for the host city may hinder the receipt of treatment, cause damage, scratches, and loss of quality of care, unlike those who live near or in the same city the center of reference[9].

In addition to these factors, the distance between home and the reference center and financial difficulties with transport and interfere with the adherence[18]. These difficulties often can aggravate the disease and compromise the health of users with hemophilia[16].

PARTICIPANT	EDUCATION	PROFESSION/ OCCUPATION	MUNICIPALITY OF RESIDENCE	LOVING RELATIONSHIP	HAS SON (A)
H1	Higher education in progress	Student	Pelotas	Single with girlfriend	Not
H2	incomplete primary education	Retired disease (working with general services)	Pelotas	Single with girlfriend	Not
H3	incomplete primary education	Retired disease (working in agriculture)	Canguçu	Married	Two daughters
H4	complete higher education	Student	Pelotas	Married	Not
H5	incomplete primary education	Retired disease (working in agriculture)	Canguçu	Married	Two daughters
H6	incomplete primary education	Retired disease (working on leather trim and agriculture)	São Lourenço do Sul	Married	A daughter and a son (stillborn)
H7	Higher education in progress	Student	big River	Single with girlfriend	Not
H8	Complete high school	Retired disease (works as a freelance)	Pelotas	Married	One daughter
H9	Higher education in progress	Student	Pelotas	Not married	Not
H10	Complete high school	Retired disease (working with general services)	big River	Divorced	One daughter
H11	incomplete primary education	Receives benefit by disease (working in agriculture)	St. Joseph North	Married	Not
H12	complete higher education	She works in the public sector	Piratini	Married	A daughter and three sons

Table 2. Characteristics of participants according to sociodemographic variables. Pelotas, Brazil, 2015 (n = 12).

Because of these reasons, it was implemented in Brazil in 1999 Household Dose Program, in which the user takes for your home doses of coagulation factor concentrate, to speed up the infusion of this, which can be performed at home or in service itself health and therefore reduce pain, hemophilic arthropathy and promote a humanized care[1]. This prevents stress due to the need for travel to the specialist service, absence from work and school, providing more time for other activities and accountability and adherence to treatment.

In Minas Gerais, a study on the reference center for the treatment of hemophilia, found that 70% of users with moderate or severe form of the disease have trained to perform the treatment in his home, which represents a good coverage by the program that seeks to include new users and family periodically[13].

As for education, it was observed that five (41.7%) had incomplete primary education, two (16.7%) completed high school, three (25%) were enrolled in higher education and only two (16.7%) had completed higher education. Among those with higher education, one (50%) was doing graduate. According to some participants, especially the older ones could not continue his studies because of various bleeding and consequences of hemophilia that resulted in absences at school. This is due to late diagnosis and treatment. In cases of bleeding, physical limitations or long hospitalizations, end up absent or leaving the work and studies, and can be fired until the job[16-18].

Regarding the profession / occupation, 4 (33.3%) were students from different areas, aged 19-28 years. A user with 40 years working in the public sector. In the age, group 39-55 years, 6 (50%) were retired and receiving benefits by disease. Previously, they worked in agriculture with general services and leather finishing, among these, one was still working as a freelance. Family income ranged from 1100.00 to 15000.00 real.

According to the presented reports, pensions and benefit occurred by hemophilic arthropathy that caused disability and pain, making it impossible to carry out their activities and stay in employment[17]. In a study of 33 users with hemophilia treated at a center of reference, the majority (72.7%) received some kind of government benefits such as free, duty-free and/or retirement moderate disability due to arthropathies and/or severe[16].

Authors of Singapore concluded by means of a study that one-fifth of people with moderately severe hemophilia A and B were unemployed, three quarters had no health insurance coverage, and about half received additional government assistance for your coagulation factor concentrate[14]. In Minas Gerais, other authors have said that more than half (51.3%) of participants in a survey received some form of social benefit, since they were considered unable to work[13].

It is essential to note that the user with hemophilia have intellectual capacity, physical and relationships as anyone else, so it is necessary to insert it socially, as in physical activity in student life and the labor market, and create opportunities means suitable for such[16-17]. With the increased availability of clotting factors, available treatments, physical activity and physical therapy, other rehabilitation measures should be encouraged in order to reduce morbidity imposed by hemophilic arthropathy, thus improving the quality of life of users[8,13,18].

Regarding marital status, the 12 participants, 7 (58.3%) were married, one (8.4%) and separated from the others (33.3%) singles. Among married and separated, they had six children. They reported the fear of transmitting hemophilia to his descendants, because they know their suffering. However, the decision to have children not influenced by its history with the disease and the transmissibility to the next generations. Participants in another study reported that the experiences with hemophilia could lead to the decision to have children in order to preserve a next child of the suffering caused by the disease[19].

Regarding family history of hemophilia ten (83.3%) had a history, including two pairs of brothers interviewed, and one of the doubles was familiar of another participant. Of the ten members with family history, nine (90.0%) reported that family died of hemophilia-related problems. For most, have family history helped in the understanding of the disease and care, despite the difficulties and the challenges of living with her. In another study, some respondents said they have learned to deal with the disease with the example of a family also affected[20].

When you have other family members with the same disease, knowledge can help in the process of understanding, behavior, and adaptation, leading some to consider as something "normal" disease in the family. However, this is not so simple, because in many cases involves anguish because of frustrating stories as familiar with several hemophiliac arthropathy, severe bleeding and deaths associated with the disease. Moreover when history, as previous generations have suffered too the prejudice of society, difficulty with diagnosis and treatment. These factors contribute to the quality of life moderate due, among other factors, difficulties in access to health care[16].

At the same time, have no family history also entails anxieties, fears and uncertainties due to the lack of prior knowledge and difficulties in dealing with situations and complications imposed by hemophilia and the lack of provision of treatment and trained professionals to treat and provide guidance adequate and safe. On the other hand, can further stimulate the search for information about the disease and treatment.

It is vital that health professionals to diagnose a person with hemophilia are cozy and guide the user and his family clearly and adequately always seeking the individual understanding, with attitudes that provide confidence and security in family restructuring process[1]. Among these professionals, the nurse plays a key role, as it constitutes a link between the service user, their family, and other professional staff[8]. It is important to have a full look, a sensitive listening, teach and guide, helping the men in the process of living with hemophilia and their resulting adaptations.

FINAL CONSIDERATIONS

It understood that describe the sociodemographic characteristics of men with hemophilia may allow health professionals, society and family understanding of who these people are. With regard to health professionals, nurses can enable the planning of actions that meet user needs with hemophilia, with the aim of promoting the improvement of care, adherence to treatment and quality of life. Also, involve and encourage everyone, patients, families, and social groups, as knowledge and understanding of the disease and its consequences, as well as the behavior and practice of integrated care.

The study presents as main limitation the limited number of participants to a better characterization. However, it is noteworthy that this study is a qualitative research. It is proposed and it expected that other researchers feel encouraged to develop studies with hemophiliacs users, looking beyond its characterization understand the subjective aspects of their experience. It considered also that the results could contribute to the identification of the care needs of these people by health professionals, especially nurses working in centers for the treatment of hemophilia, and provide the opportunity for understanding of the management of disease and improving the quality of life of these people.

CARACTERÍSTICAS SOCIODEMOGRÁFICAS DE HOMENS COM HEMOFILIA NO SUL DO BRASIL

RESUMO

Objetivou-se descrever as características sociodemográficas de homens com hemofilia em acompanhamento por um Hemocentro Regional situado no Sul do Rio Grande do Sul, Brasil. Trata-se de um estudo qualitativo e descritivo, cuja coleta de dados ocorreu entre junho de 2014 e novembro de 2015. Aplicou-se a técnica de entrevista intensiva e notas de campo e a análise dos dados foi do tipo descritiva. Participaram 12 homens com a forma grave da hemofilia e idade entre 19 e 55 anos, a maioria com hemofilia A, são casados, residentes em municípios da zona Sul do Rio Grande do Sul, com doenças infectocontagiosas, sequelas articulares e histórico de antecedentes familiares de hemofilia que evoluíram para o óbito, e metade deles com filhos e aposentados por invalidez. Conclui-se que descrever as características sociodemográficas dos homens com hemofilia pode aproximar os profissionais de saúde, sociedade e família acerca de quem eles são, além de promover o conhecimento sobre a doença e suas consequências sociais e fomentar uma prática de cuidados voltada para as necessidades de saúde e promoção da qualidade de vida.

Palavras-chave: Hemofilia A. Hemofilia B. Doença crônica. Masculino. Enfermagem.

CARACTERÍSTICAS SOCIODEMOGRÁFICAS DE HOMBRES CON HEMOFILIA EN EL SUR DE BRASIL

RESUMEN

el objetivo fue describir las características sociodemográficas de hombres con hemofilia acompañados por un Hemocentro Regional ubicado en el sur de Rio Grande do Sul, Brasil. Se trata de un estudio cualitativo y descriptivo, cuya recolección de datos ocurrió entre junio de 2014 y noviembre de 2015. Se utilizó la técnica de entrevista intensiva y notas de campo y el análisis de los datos fue del tipo descriptivo. Participaron 12 hombres con el tipo grave de hemofilia y edad entre 19 y 55 años, la mayoría con hemofilia A, casados, viven en municipios de la región sur de Rio Grande do Sul, con enfermedades infectocontagiosas, secuelas articulares e histórico de antecedentes familiares de hemofilia que evolucionaron para el óbito, la mitad de ellos con hijos y jubilados por invalidez. Se concluye que describir las características sociodemográficas de los hombres con hemofilia puede aproximar a los profesionales de salud, la sociedad y a la familia acerca de quiénes ellos son, además de promover el conocimiento sobre la enfermedad y sus consecuencias sociales y fomentar una práctica de cuidados dirigida para las necesidades de salud y la promoción de la calidad de vida.

Palabras clave: Hemofilia A. Hemofilia B. Enfermedad crónica. Masculino. Enfermería.

REFERENCES

1. Brasil. Ministério da Saúde. Secretaria de Atenção à Saúde. Departamento de Atenção Especializada e Temática. Manual de hemofilia. Brasília: Ministério da Saúde; 2015 [citado em 2018 Nov]; 80p. Disponível em: http://bvsms.saude.gov.br/bvs/publicacoes/manual_hemofilia_2ed.pdf.

2. World Federation of Hemophilia. Report on the Annual Global Survey 2016. Canada: World Federation of Hemophilia; 2017 [citado em 2018 Nov.]; 80p. Disponível em: https://www1.wfh.org/publication/files/pdf-1690.pdf.

3. Brasil. Ministério da Saúde. Secretaria de Atenção à Saúde. Departamento de Atenção Especializada e Temática. Perfil das coagulopatias hereditárias no Brasil: 2015. Brasília: Ministério da Saúde; 2017[citado em 2018 Nov]; 68p. Disponível em: http://bvsms.saude.gov.br/bvs/publicacoes/perfil_coagulopatias_hereditarias_brasil_2015.pdf.

4. Olsson A, Hellgren M, Berntorp E, Baghaei F. Association between bleeding tendency and health-related quality of life in carriers of moderate and severe hemophilia. Haemophilia [on-line]. 2015[citado em 2018 nov.]; 21(6):742-6. doi: https://doi.org/10.1111/hae.12796.

5. Castillo González D. La hemofilia: situación actual en Cuba y perspectivas. Rev. Cubana Hematol. Inmunol. Hemoter. [On-line]. 2013 [citado em 2015 nov.]; 29(2):112-3. Available from: http://scielo.sld.cu/scielo.php?script=sci_arttext&pid=S0864-02892013000200001&lng=es&nrm=iso.

6. Kodra Y, Cavazza M, Schieppati A, De Santis M, Armeni P, Arcieri

R, et al. The social burden and quality of life of patients with hemophilia in Italy. Blood Transfus [on-line]. 2014[citado em 2018 Nov], 12(Suppl. 3):s567-75. doi: https://doi.org/10.2450/2014.0042-14s.

7. Guh S, Grosse SD, Ullman M, Soucie JM. Accounting for differences in healthcare utilization and expenditures among US males with hemophilia by type of health insurance. Haemophilia [on-line]. 2017 [citado em 2018 Nov]; 23(2): e147-51. doi: https://doi.org/10.1111/hae.13161.

8. Santos RS, Figueirôa GR, Machado BA, Mamede CA, Gois LML, Junior CMS. Frequency of postural alterations in hemophiliacs. J Phys Res [on-line]. 2018[citado em 2018 Nov.]; 8(1):24-36. doi: http://dx.doi.org/10.17267/2238-2704rpf.v8i1.1601.

9. Veloso HH, Silva, NA, Araújo JSM. Perfil epidemiológico dos portadores de hemofilia do hemocentro da Paraíba. Rev. Odontol. Bras. Central [on-line]. 2013 [citado em 2018 Nov]; 21(61):103-9. Disponível em: http://www.robrac.org.br/seer/index.php/ROBRAC/article/view/769/687.

10. Santos BP, Feijó AM, Viegas AC, Schwartz E, Lise F. Classificação das pesquisas. In: Lise F, Souza, BM, Schwartz E, Garcia FRM. Etapas da construção científica: da curiosidade acadêmica à publicação dos resultados. Pelotas: UFPel; 2018 [citado em 2018 Nov]; p.61-74. Disponível em: http://guaiaca.ufpel.edu.br:8080/handle/prefix/4171.

11. Brasil. Ministério da Saúde. Conselho Nacional de Saúde. Resolução nº 510, de 7 de abril de 2016. Diário Oficial da República Federativa do Brasil, Brasília, DF, 24 maio 2016. Seção 1 [citado em 2018 Nov]. Disponível em: http://conselho.saude.gov.br/resolucoes/2016/reso510.pdf.

12. Rocha P, Carvalho M, Lopes M, Araújo F. Costs and utilization of treatment in patients with hemophilia. BMC Health Serv. Res [on-line]. 2015[citado em 2018 Nov]; 15(1):484-90. doi: https://doi.org/10.1186/s12913-015-1134-3.

13. Ferreira AA, Leite ICG, Bustamante-Teixeira MT, Corrêa CSL, Cruz DT, Rodrigues DOW, et al. Health-related quality of life in hemophilia: results of the Hemophilia-Specific Quality of Life Index (Haem-a-Qol) at a Brazilian blood center. Rev Bras Hematol Hemoter [on-line]. 2013 [citado em 2018 Nov], 35(5):314-8. doi: http://dx.doi.org/10.5581/1516-8484.20130108.

14. Ng HJ, Lam J, Koh PL, Ho L, Lim CY, Akhbar Ali M, et al. A comprehensive study of current hemophilia care and outcomes in Singapore. Haemophilia [on-line]. 2015[citado em 2018 Nov]; 21(5): e428-31. doi: https://doi.org/10.1111/hae.12729.

15. García-Chávez J, Majluf-Cruz A. Hemofilia. Gac Med Mex [on-line]. 2013[citado em 2018 Nov];149(3):308-21. Disponível em: http://www.medigraphic.com/pdfs/gaceta/gm-2013/gm133j.pdf.

16. Souza JG, Vieira RTF, Duarte ARP, Figueira MCS, Jacob LMS, De Melo MC. Qualidade de vida dos portadores de hemofilia a em um Hemocentro na região amazônica. Rev. Intellectus [on-line]. 2018[citado em 2018 Nov]; 44(1): 33-45. Disponível em: http://www.revistaintellectus.com.br/DownloadArtigo.ashx?codigo=913.

17. Garbin LM, Carvalho EC, Canini SRMS, Dantas RAS. Avaliação da qualidade de vida relacionada à saúde em pacientes portadores de hemofilia. Cienc. Cuid Saúde [on-line]. 2007 [citado em 2018 Nov.]; 6(2):197-205. doi: http://dx.doi.org/10.4025/cienccuidsaude.v6i2.4153.

18. Targino Junior MA, Leite Filho MAA, Montenegro RC, Barbosa EL. Antropometria e força muscular de indivíduos hemofílicos da cidade de João Pessoa-PB. RBPFEX [on-line]. 2017 [citado em 2018 Nov]; 11(69):743-7. Disponível em: http://www.rbpfex.com.br/index.php/rbpfex/article/view/1261/977.

19. Apolinário LA, Rodrigues LR. Mães de crianças e adolescentes hemofílicos e suas concepções sobre maternidade. REAS [on-line]. 2013 [citado em 201 Nov]; 2(3):42-52. doi: https://doi.org/10.18554/.

20. Lindsay K, Gow P, Vanderpyl J, Logo P, Dalbeth N. The Experience and Impact of Living with Gout: A Study of Men with Chronic Gout Using a Qualitative Grounded Theory Approach. J Clín. Rheumatol [on-line]. 2011[citado em 2018 Nov]; 17(1):1-6. doi: https://doi.org/10.1097/RHU.0b013e318204a8f9.

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