79 - QUALITY OF LIFE IN HEMOPHILIACS: A STUDY IN INTERNATIONAL DATABASES

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INTRODUCTION

Hemophilia is a hereditary disease, hemorrhagic, predominantly due to deficiencies quantitative in the biosynthesis of factors or clotting proteins, or molecular defects (qualitative deficiencies of factors VIII and IX) (VILLAÇA, RAM, D'AMICO, 2001).

The incidence of hemophilia in the various ethnic groups is approximately 1:10,000 births. In 85% of cases are caused by deficiency of factor VIII, being called hemophilia A or classic hemophilia and approximately 15% of cases are caused by deficiency of factor IX (hemophilia B) (BRAZIL, 2008).

In most cases, hemophilia is transmitted to the child by the carrier mother , which is usually asymptomatic. However, about 30% of cases of hemophilia result from new mutations, ie, occurs in the absence of cases in other family members (ROIZEN, 2000).

Clinically, hemophilia A and B are similar and the diagnosis is made by measuring the activity of factors VIII and IX coagulation. The classification of hemophilia varies depending on the level antigen (Ag) or coagulant activity (C) of the deficient factor, being the normal level defined as 1 IU / ml or 100%, respectively (BRAZIL, 2008).

According to consensus of the International Society of Thrombosis and Haemostasis, it is recommended to classify as severe hemophilia, moderate and mild if the Ag or C is <0.01 IU / mL and / or <1%, 0,01-0,05 IU / mL and / or 1-5% and> 0.05 to <0.40 IU / mL and / or> 5 to <40%, respectively (GARBIN et al, 2007).

From our experience as health professionals, assisting patients with hemophilia and realizing the impact of quality of life in haemophilia, aroused the interest to perform a search in the literature about the topic.

Therefore, we believe that the systematic survey of publications that addresses quality of life in hemophilic patients is very important for professionals to know the national and international reality and the way of how this object of study is being published in order to provide the customers with a holistic care, aiming to promote the health of the patient with hemophilia.

Given the above, emerged the following questions: How has occurred the publication involving "quality of life" and "hemophilia" in the databases of the Virtual Health Library / Regional Library of Medicine (BVS / BIREME)? What are the years that most published studies? What types of studies and approaches used? How available? What are the most researched topics? And what are the pioneers in researching this topic?

To answer these questions, was developed the following goal: to characterize the scientific literature on quality of life of hemophiliacs in the databases of the BVS / BIREME (LILACS, MEDLINE, SCIELO, ADOLEC and MEDCARIB) according as the year of publication (2003 to April 2009), type of study and approach, form of publication (abstract and complete), linked to thematic categories, language (English, Portuguese and Spanish) and country of publication.

RESULTS AND DISCUSSION

The research is an exploratory, descriptive and prospective with data and a quantitative approach based the data from BVS / BIREME (LILACS, MEDLINE, SCIELO, ADOLEC and MEDCARIB).

Data collection was performed during the month of May 2009, from a vast literature in electronic databases listed above. The descriptors used for data collection were: "hemophilia" and "quality of life", according to the classification of the descriptors in the Health Sciences (DECS).

The criteria for inclusion of articles for this literature review points to studies on the issue of quality of life in patients with hemophilia, published between the years 2003 to April 2009 in English, Portuguese, Spanish, in the form of full-text or abstract. The exclusion criteria have focused for the studies that did not respond to questioning and that were published in more than one database. The cohort of the study period is justified for ensuring the timeliness of the data, focusing on trends in analyzed research.

During the collection, there were a total of 74 articles distributed as follows: 01 in LILACS, 72 in MEDLINE and 1 in ADOLEC . In SCIELO and MEDCARIB did not find articles that met the study. Data were collected using a structured form, covering issues consistent with the research proposal, these were also recorded and analyzed in Microsoft Excel spreadsheets 2007, using descriptive statistics and presented as tables and graphs.

Table 1 summarizes the quantity of articles of the type of study and approach, and the form of publication in the databases examined.

Table 1. Distribution of articles surveyed on quality of life in haemophilia in the databases MEDLINE, LILACS and ADOLEC as the type of study, approach and form of publication. Natal / RN, 2009.

VARIABLES	MEDLINE		LILACS		ADOLEC		TOTAL	
	N	%	N	%	N	%	N	%
Type of study								
Descriptive	26	35,1	1	1,4	1	1,4	28	37,8
Theoretical review	20	27,0	0	0,0	0	0,0	20	27,0
Cross-sectional study	10	13,5	0	0,0	0	0,0	10	13,5
Experimental study	7	9,5	0	0,0	0	0,0	7	9,5
Case-control	5	6,8	0	0,0	0	0,0	5	6,8
Cohort study	2	2,7	0	0,0	0	0,0	2	2,7
Case Study	2	2,7	0	0,0	0	0,0	2	2,7
Type of approach								
Quantitative	39	52,7	1	1,4	1	1,4	41	55,4
Qualitative	30	40,5	0	0,0	0	0,0	30	40,5
Quali-quantitative	3	4,1	0	0,0	0	0,0	3	4,1
Form of publication								
Abstract	70	94,6	0	0,0	1	1,4	71	95,9
Full	2	2,7	1	1,4	0	0,0	3	4,1
Total	**72**	**97,3**	**1**	**1,4**	**1**	**1,4**	**74**	**100,0**

As can be seen in Table 1, most of the papers surveyed used the descriptive study (37.8%), with a quantitative approach (55.4%) and published in an abstract form (95.9%).

Regarding the type of study, dominated the descriptive, which observes, records, analyzes and correlates the facts of the physical world occurring in the universe perceived by man, describing their structure and function without interference from the researcher. Find out exactly how often the phenomenon occurs, their relationship and connection with others (CRUZ; RIBEIRO, 2004).

About the type approach, quantitative research was the most used. The quantitative paradigm, hegemonic in biomedical research, uses methods from the physical sciences, epidemiology and statistics. It is characterized by the adoption of deductive methods and seeks objectivity, validity and reliability (SANTOS, 1999).

The quantitative research is characterized by the performance levels of reality and presenting as goals the identification and presentation of data, indicators and trends observable. This kind of research shows it is generally appropriate when there is the possibility of collecting quantifiable measures of variables and inferences from samples of a population (CARMO; FERREIRA, 1998).

As regards the form of publication, one realizes that most of the articles were available only as a abstract, which did not preclude the answers of the questions this work. However, further information shall be prejudiced, since they would need the availability of full text articles, which represents a gap found during this research.

when about the years of publications, we identified that the electronic scientific literature about the quality of life in hemophilia had a great expression in the years 2008 (27.0%) and 2004 (19.0%). As the topic is very current and important, we realize that the studies have staying in evidence over the years, but there is a clear decrease between 2005 and 2007. Until April 2009 was not found research on the subject available in the databases investigated.

Regarding the language in which the works were published, 97.3% were available in English, Portuguese 1.4% and 1.4% in Spanish.

Although the English language be considered as universal language, most of the people of the world does not speak or read English,since they are in underdeveloped countries and don´t has access to another language, that is not spoken in your country. This fact restricts access to information in developing countries. However, the availability of data in English ensures universality of information to the community that dominates that language.

Taking into account the country where the research was published, there have been: England (68.9%), United States of America - USA (14.9%), Germany (5.4%), Italy (2.7 %), Venezuela, Spain, Poland, New Zealand, Japan and France relied on 1.4% each.

We can see that the developed countries, especially England and the U.S.A., are major investors in research on quality of life in hemophilic patients, this accounts for the availability of jobs in English, since these countries have the language as official language.

However, note that developing countries just do not invest in research on the subject, only Venezuela has 1.4% of the studies found. Brazil has no publications in the area linked the databases searched.

For better understanding of the studies analyzed, the articles were grouped in themes. In Table 2, are arranged such areas and their relative and absolute values as the electronic databases.

Table 2. Distribution of articles surveyed about quality of life and hemophilia in the databases MEDLINE, LILACS and ADOLEC as the thematic framed. Natal / RN, 2009.

THEMATIC	MEDLINE		LILACS		ADOLEC		TOTAL	
	N	%	N	%	N	%	N	%
Psychological adaptation	22	29,7	1	1,4	0	0,0	23	31,1
Several treatments	15	20,3	0	0,0	0	0,0	15	20,3
Child and adolescent	13	17,6	0	0,0	1	1,4	14	18,9
Treatment with the use of factor VIII	13	17,6	0	0,0	0	0,0	13	17,6
Elderly	5	6,8	0	0,0	0	0,0	5	6,8
Epidemiology	4	5,4	0	0,0	0	0,0	4	5,4
Total	**72**	**97,3**	**1**	**1,4**	**1**	**1,4**	**74**	**100,0**

According to Table 2, we find that most of the works were focused in psychological adaptation in relation to quality of life in hemophilic patients (31.1%), followed by several treatments (20.3%), studies that dealing with the issue in children and adolescents (18.9%), treatment with the use of factor VIII (17.6%), studies in elderly (6.8%) and epidemiology of the disease (5.4%).

A study realized in the blood center in Campinas, SP, showed that most of those interviewed considers hemophilia as a disease that interferes with social life (60%), affective (61%), professional (67%) and psychological wellbeing (80%) of their carriers (CAIO, 2001).

Arranz (2004) argues that a profound effect on self-esteem of these individuals may explain the high proportion of negative feelings (depression, anxiety, insecurity, feelings of injustice, anger and fear) in relation to self-diagnose hemophilia.

Although some individuals, position yourself with ease in the face of their situation, demonstrating a sense of apparent normality, it is difficult to rule out the possibility of denial, a psychological mechanism to protect the ego (CAIO, 2001).

Depression and anxiety are the most frequent psychological changes associated with hemophilia in the literature. Patients with hemophilia have a personality profile in which the predominant inhibition, dependence and immaturity (ARRANZ, 2004).

The treatment of haemophilia is based mainly on the replacement of the deficient clotting factor (factor VIII concentrates or IX), usually obtained from human plasma. Recombinant concentrates, ie developed by modern molecular biology techniques and highly purified, are also used in several countries (MARTINEZ, 2004).

Products derived from human plasma (blood derivatives) are the most commonly used to treat hemophilia. New diagnostic techniques, viral inactivation and purification of blood products has become very safe products, but there is always the fear of transmission of viruses and other infectious agents through of products sourced from human plasma, as happened with the HIV virus (AIDS) and the hepatitis viruses B and C (MARTINEZ, 2004).

CONCLUSIONS

Most of the articles surveyed were used in a descriptive quantitative approach and published in abstract form. About the year, there were strong expression in the years 2008 and 2004, decreased between 2005 and 2007.

Regarding the language in which the works were published, most were available in English. This shows downside regarding the difficulty of the general community of developing countries have access to these publications. But there is the advantage of these works are available in a universal language, ensuring the dissemination to the scientific community.

Taking into account the country where the research was published, the main sites were England, United States of America (USA), Germany and Italy. Venezuela appears as the only developing country to publish article on the subject and Brazil did not include studies in the databases investigated. The lack of studies in Brazil suggest a large knowledge gap to be filled in the Country. This fact should be reviewed by researchers in the field of health, to develop more studies and publish them to knowledge of the academic and general.

Finally, on the themes more addressed in quality of life of hemophiliacs were psychological adaptation, use of various treatments, studies in child and adolescent treatment and use of factor VIII.

We conclude that, to face the day-to-day in the normal way, it is recommended to hemophiliacs, the practice of regular exercise (except the high-impact sports) such as swimming, encouraged to increase the inclusion of patients in activities social subjects.

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QUALITY OF LIFE IN HEMOPHILIACS: A STUDY IN INTERNATIONAL DATABASES

ABSTRACT

Hemophilia is a hereditary disease, hemorrhagic, predominantly, due the quantitative to deficiencies in the biosynthesis of factors or clotting proteins, or molecular defects (qualitative deficiencies of factors VIII and IX). This study aimed to characterize the scientific literature on quality of life of hemophiliacs in the databases of the BVS / BIREME (LILACS, MEDLINE, SCIELO, ADOLEC and MEDCARIB) according as the year of publication (2003 to April 2009), type of study and approach, form of publication (abstract and complete), linked to thematic categories, language (English, Portuguese and Spanish) and country of publication. The research is exploratory and descriptive study with prospective data and quantitative approach, carried out in electronic databases available in the BVS / BIREME. Data collection was performed in May/2009, with the use of descriptors "hemophilia" and "quality of life". During the collection, there were found 74 articles: 01 in LILACS, 72 in MEDLINE and ADOLEC in 1. In SCIELO and MEDCARIB no articles were found. Most of the articles surveyed used the descriptive study (37.8%), with a quantitative approach (55.4%), published in an abstract form (95.9%) in the years 2008 (27.0%) and 2004 (19.0%), in english (97.3%) developed in England (68.9%) and USA (14.9%) about psychological adaptation in relation to quality of life in hemophilic patients (31 , 1%), various treatments (20.3%). To avoid psychological problems and improve quality of life is necessary that the patients and their families receive more information about the disease as well as the population and health professionals. Through national campaigns can improve the quality of medical services and encourage patient adherence to treatment.

KEYWORDS: Hemophilia, quality of life, publication, nursing

QUALITE DE VIE CHEZ LES HEMOPHILES: EXAMEN D'INTEGRATION DANS BASES DE DONNEES ELECTRONIQUES

L'hémophilie est une maladie héréditaire, hémorragique, principalement en raison de lacunes dans la biosynthèse des facteurs quantitatifs ou des protéines de la coagulation, ou des défauts moléculaires (déficiences qualitatives de facteurs VIII et IX). Cette étude vise à caractériser la littérature scientifique sur la qualité de vie des hémophiles dans les bases de la BVS / BIREME (LILACS, MEDLINE, SciELO, PubMed et MEDCARIB) comme l'année de publication (de 2003 à avril 2009), le type de étude et d'approche, une forme de publication (résumé et complet), liée à des catégories thématiques, langue (anglais, portugais et espagnol) et pays de publication. L'étude de la recherche est exploratoire et descriptive des données prospectives et approche

quantitative, réalisée dans des bases de données électroniques disponibles dans la BVS / BIREME. La collecte des données a été réalisée en Mai/2009, avec l'utilisation de descripteurs «hémophilie» et la «qualité de vie." Lors de la collecte, il y avait 74 articles: 01 LILACS, 72 dans MEDLINE et PubMed en 1. En SCIELO et MEDCARIB aucun article n'a été trouvé. La plupart des articles interrogées utilisaient l'étude descriptive (37,8%), avec une approche quantitative (55,4%), publiée dans une forme abstraite (95,9%) dans les années 2008 (27,0%) et 2004 (19,0%), anglais (97,3%) ont développé en Angleterre (68,9%) et USA (14,9%) sur l'adaptation psychologique en matière de qualité de vie des patients hémophiles (31,1%), divers traitements (20,3%). Pour éviter des problèmes psychologiques et d'améliorer la qualité de vie est nécessaire pour les patients et leurs familles bénéficient de plus amples renseignements sur la maladie et la population ainsi que les professionnels de santé. Par des campagnes nationales peuvent améliorer la qualité des services médicaux et d'encourager l'adhésion des patients au traitement.

MOTS-CLÉS: Hémophilie, Qualité de vie; Publications; Soins infirmiers.

CALIDAD DE VIDA EN LOS HEMOFÍLICOS: UNA REVISIÓN INTEGRADORA DE LAS BASES DE DATOS ELECTRÓNICAS

La hemofilia es una enfermedad hereditaria hemorrágica, debido principalmente a deficiencias en la biosíntesis de los factores cuantitativos o de proteínas de la coagulación, o defectos moleculares (deficiencias cualitativas de los factores VIII y IX). Este estudio tuvo como objetivo caracterizar la literatura científica sobre la calidad de vida de los hemofílicos en las bases de datos de la BVS / BIREME (LILACS, MEDLINE, SciELO, PubMed y MEDCARIB) por el año de publicación (2003 a abril de 2009), el tipo de estudio y enfoque, una forma de publicación (resumen y completa), vinculado a las categorías temáticas, idiomas (Inglés, portugués y español) y país de publicación. El estudio de la investigación exploratoria y descriptiva con datos cuantitativos y el enfoque prospectivo, realizado en bases de datos electrónicas disponibles en la BVS / BIREME. La recopilación de datos se realizó en Mayo/2009, con el uso de los descriptores "hemofilia" y "calidad de vida". Durante la busca, se ha encontrado a 74 artículos: 01 en LILACS, 72 en MEDLINE y 1 en PubMed. En SciELO y MEDCARIB no fueron encontrados artículos. La mayoría de los artículos encuestadas utilizaban el estudio descriptivo (37,8%), con un enfoque cuantitativo (55,4%), publicada en una forma abstracta (95,9%) en los años 2008 (27,0%) y 2004 (19,0%), Inglés (97,3%) se desarrolló en Inglaterra (68,9%) y EUA. (14,9%) en la adaptación psicológica en relación con la calidad de vida en pacientes hemofílicos (31 , 1%), los diversos tratamientos (20,3%). Para evitar los problemas psicológicos y mejorar la calidad de vida es necesario para los pacientes y sus familias reciben más información sobre la enfermedad y la población, así como profesionales de la salud. A través de campañas nacionales pueden mejorar la calidad de los servicios médicos y fomentar la adhesión del paciente al tratamiento.

PALABRAS CLAVE: Hemofilia; Calidad de vida; Publicaciones; Enfermería.

QUALIDADE DE VIDA EM HEMOFÍLICOS: UMA REVISÃO INTEGRATIVA EM BASES DE DADOS ELETRÔNICAS

RESUMO

A hemofilia é uma doença hereditária, hemorrágica, predominantemente, decorrente de deficiências quantitativas na biossíntese de fatores ou proteínas de coagulação, ou de defeitos moleculares (deficiências qualitativas dos fatores VIII e IX). O objetivo desse trabalho é caracterizar a produção científica sobre qualidade de vida em hemofílicos nas bases de dados da BVS/BIREME (LILACS, MEDLINE, SCIELO, ADOLEC E MEDCARIB), conforme o ano de publicação (2003 a abril de 2009), tipo de estudo e abordagem, forma de publicação (resumo e completo), categorias vinculadas a temática, idioma (português, inglês e espanhol) e país de publicação. A pesquisa é do tipo exploratório-descritivo, com dados prospectivos e abordagem quantitativa, realizada nas bases de dados eletrônicas disponíveis na BVS/BIREME. A coleta de dados foi realizada em maio/2009, com a utilização dos descritores “hemofilia” e “qualidade de vida”. Durante a coleta, foram encontrados 74 artigos: 01 na LILACS, 72 na MEDLINE e 1 na ADOLEC. No SCIELO e na MEDCARIB não foram achados artigos. A maioria dos artigos pesquisados utilizou-se do estudo descritivo (37,8%), com abordagem quantitativa (55,4%), publicação na forma de resumo (95,9%), nos anos de 2008 (27,0%) e 2004 (19,0%), em inglês (97,3%), elaborados na Inglaterra (68,9%) e EUA (14,9%), sobre adaptação psicológica com relação a qualidade de vida em pacientes hemofílicos (31,1%), tratamentos diversos (20,3%). Para evitar transtornos psicológicos e melhorar qualidade de vida faz-se necessário que os pacientes e seus familiares recebam mais informações sobre a doença, assim como a população e os próprios profissionais de saúde. Através de campanhas nacionais pode-se melhorar a qualidade dos serviços médicos e estimular a adesão dos pacientes ao tratamento.

PALAVRAS-CHAVE: Hemofilia; Qualidade de vida; Publicações; Enfermagem.

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