INTEGRATIVE LITERATURE REVIEW ARTICLE

SELF-CARE OF THE HEMOPHILIC CHILD*

O AUTOCUIDADO DO ESCOLAR HEMOFÍLICO

EL AUTOCUIDADO DEL NIÑO CON HEMOFILIA

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ABSTRACT

Objective: to verify how children with hemophilia A and B perform self-care. **Method:** this is a bibliographic, descriptive, integrative literature review study. We searched publications from 2013 to 2018 in the databases BDENF, LILACS, MEDLINE and SciELO Virtual Library. Data were analyzed descriptively from the establishment of categories. **Results:** five articles respond to the objective studied. After the analysis, the studies were categorized and the benefits of empowering the patient through self-management from childhood and the need to understand the chronic disease process were promoted, promoting effective self-management through pre-established routines. **Conclusion:** it is clear that there is a shortage of articles that address the Brazilian reality, making it difficult to understand the national scenario. The need for children to understand the disease and generate autonomy in the management of their self-care was identified. **Descriptors:** Nurses, Pediatric; Self Care; Hemophilia A; Hemophilia B; Hematology.

RESUMO

Objetivo: verificar como escolares com hemofilia A e B realizam o autocuidado. **Método:** trata-se de estudo bibliográfico, descritivo, tipo revisão integrativa da literatura. Buscou-se publicações entre 2013 à 2018 nas bases de dados BDENF, LILACS, MEDLINE e Biblioteca Virtual SciELO. Analisaram-se os dados de forma descritiva a partir do estabelecimento de categorias. **Resultados:** informam-se que cinco artigos respondem ao objetivo estudado. Realizou-se após a análise a categorização dos estudos e se observaram os benefícios de empoderar o paciente por meio do autogerenciamento desde a infância e a necessidade de compreender o processo de doença crônica, promovendo um autogerenciamento eficaz por meio de rotinas pré-estabelecidas. **Conclusão:** percebe-se que há escassez de artigos que abordem a realidade brasileira, dificultando a compreensão do cenário nacional. Identificou-se a necessidade do escolar compreender a doença e gerar autonomia no gerenciamento do seu autocuidado. **Descritores:** Enfermagem Pediátrica; Autocuidado; Hemofilia A; Hemofilia B; Hematologia.

RESUMEN

Objetivo: verificar cómo los niños con hemofilia A y B realizan el autocuidado. **Método:** este es un estudio de revisión bibliográfica, descriptivo, integrador de literatura. Se realizaron búsquedas en las publicaciones de 2013 a 2018 en las bases de datos BDENF, LILACS, MEDLINE y la Biblioteca Virtual SciELO. Los datos se analizaron descriptivamente a partir del establecimiento de categorías. **Resultados:** cinco artículos responden al objetivo estudiado. Después del análisis, se categorizaron los estudios y se promovieron los beneficios de empoderar al paciente a través del autocontrol desde la infancia y la necesidad de comprender el proceso de las enfermedades crónicas, promoviendo el autocontrol efectivo a través de rutinas preestablecidas. **Conclusión:** está claro que hay una escasez de artículos que aborden la realidad brasileña, lo que dificulta la comprensión del escenario nacional. Se identificó la necesidad de que los niños entiendan la enfermedad y generen autonomía en el manejo de su autocuidado. **Descriptores:** Enfermería Pediátrica; Autocuidado; Hemofilia A; Hemofilia B; Hematología.

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INTRODUCTION

Nursing is known to be active in several specialties and, among its biases, there is care for patients with hemophilia A and B. It is a disease, mostly hereditary, recessive and congenital, related to the X chromosome and characterized by hindering the blood clotting process, usually due to deficiency of factors VIII and IX, respectively.¹ It is added that it can present mild, moderate or severe that can result in lifelong obstacles, especially in childhood.

Since 2011, the Ministry of Health of Brazil has promoted a series of advances in the policy of care for patients with hereditary coagulopathies that positively impacted quality of life, such as the implementation of immunotolerance and primary prophylaxis treatments. In addition to these, there are also those who use genetic engineering to obtain the factors that, being synthesized in the laboratory, minimize the risks of the hemophilic organism to create antibodies.²

It is pointed out that chronic disease accompanies the individual throughout their life, going through acute, worsening or significant improvement phases.³ The bearer must develop, throughout his life, the ability to self-care. You must understand your limitations and work for your independence. According to Dorothea Orem's Theory of Self-Care, developed in 1971, the nurse must identify, with the client, the deficits of their individual self-care needs and seek to develop potentials for the practice, doing what the patient a priori , cannot do.⁴

Five methods that interfere with nursing care are proposed by Orem and should be evaluated by nurses in order to better conduct their care, namely: 1) assessing self-care skills; 2) self-care actions; 3) identify the demands of therapeutic self-care; 4) understand the self-care deficit and 5) enable the nursing staff to provide complete care. It is noteworthy that it is the nurse's role to teach and guide the patient, regardless of their age group.⁴ Therefore, the work is justified by the need for the nurse to encourage and create devices for the self-care management of the studied public, aiming at the development of its capabilities so that it assumes co-responsibility for its.

It develops the responsibility to self-provide in everyday life through a spontaneous learning process aided by intellectual curiosity, obtaining new information, supervising others and also by the experience gained in performing basic self-care measures.⁴ Empowered if the child is daily self-care, creating a system where it is possible, within its limitations, to self-manage. By this attitude, the child's cognitive and social development is stimulated, allowing his caregiver

to be less burdened with the responsibility of his integral care,⁵ in addition, the child with chronic disease learns about its limitations and adequacy. To the reality in which it is inserted, providing a responsible conscience about its space in the social environment.

OBJECTIVE

- To verify how children with hemophilia A and B perform self-care.

METHOD

This is a bibliographic, descriptive, integrative literature review (RIL) study. The ideal question was to answer the guiding question: "According to the current literature, how do children with hemophilia A and B perform self-care?". It consists of a six-step RIL, which dictate an elaborated and validated research protocol, as follows: 1) elaboration of the guiding question; 2) definition of inclusion and exclusion criteria; 3) categorization of studies, highlighting the commonalities between them; 4) critical discussion of the research findings; 5) interpretation of the results found and 6) disclosure of relevant information for the practice of evidence-based nursing care.⁶

The search was conducted from October to November 2018 by two independent reviewers, including the Nursing Databases (BDENF), Latin American and Caribbean Health Sciences Literature (LILACS) and Medical Literature Analysis and Retrieval System Online (MEDLINE) and the Scientific Electronic Library Online Virtual Library (SciELO). The Virtual Health Library (VHL) Health Science Descriptors (DeCS) were consulted. The following health descriptors were listed in Portuguese, English and Spanish, namely: "Pediatric Nursing"; "Self-care"; "Hemophilia A"; "Hemophilia B"; Pediatric nursing; Self-care; "Hemophilia A"; "Hemophilia B"; "Pediatric infirmary"; "Self-care"; "Hemophilia A" and "hemophilia B" using the boolean operator and.

The age range chosen was based on Jean Piaget's Cognitive Theory,⁷ which allows us to understand the cognitive development of the human being from the observation of children. For the purposes of this study, the "concrete operative" stage was used, which comprises from seven to eleven years of age.⁸ This period is responsible for marking several physiological and behavioral changes in the child that become more costly when the child is affected for chronic disease. It is revealed that the ages present in the stages elucidated by Piaget are only average, and may vary among individuals, being closely linked to the social environment and the degree of cognitive development of each child.⁹

Thus, the following inclusion criteria were employed: full texts available in Portuguese,

English and Spanish; articles that included the chosen age range and publications from 2013 to 2018. Duplicate articles that did not meet the proposed objective were excluded.

There were 2542 articles that were carefully read the titles and their respective summaries. 861 articles that were in disagreement with the study proposal were excluded, and 45 articles were separated for full-text reading; Of these, only five respond to the stated objective. Figure 1 illustrates the methodological treatment containing the simplified process of identification, selection, eligibility and inclusion of articles, with their respective numbers.

The Preferred Reporting Items for Systematic Reviews and Meta-Analyzes (PRISMA)¹⁰ recommendations were adopted to improve the review report, the Critical Appraisal Skills Program¹¹, to highlight the methodological quality of the articles, and to To evaluate the level of scientific evidence of the articles, they were classified by the level of evidence and the degree of recommendation respectively, being one as 5, three of them as 4 and one as 3; Regarding the years of publication, they occurred in 2013, 2015, 2016, 2017 and 2018.

Figure 1. Flowchart of study selection according to Preferred Reporting Items for Systematic Reviews and Meta-Analyzes (PRISMA 2009). Rio de Janeiro (RJ), Brazil, 2018.

RESULTS

All articles composing this review were indexed in the MEDLINE database. Moreover, there was a shortage in scientific production on the theme explored, especially in the national scenario, which highlights the need for deepening and making publications that discuss the self-care of

the Brazilian hemophilic school. It is noted that the country with the highest production was the United States of America (USA), with two articles, followed by Canada, the United Kingdom and India, each with one article respectively. These articles were classified by level of evidence and degree of recommendation as shown in Figure 2.

Author	Title	Year/Country	N.E	Objective	Study outline
Kachooei AR, Badiei Z, Zandinezhad ME ¹²	<i>Vicious cycle of multiple invasive treatments in a hemophilic inhibitor positive child with resistant knee flexion contracture, a case report.</i>	2013 USA	5	To describe a repetitive hemarthrosis injury in the left knee of an eight-year-old child with severe hemophilia A .	Case Study / Qualitative Study
Khair K, Meerabeau L, Gibson F ¹³	<i>Self-management and skills acquisition in boys with haemophilia</i>	2015 United Kingdom	4	Understand self-management of hemophilia from the standpoint of a 21st century child where intensive prophylactic therapy is administered from early childhood .	Qualitative study
Bérubé S, Mouillard F, Amesse C, Sultan S ¹⁴	<i>Motivational techniques to improve self-care in hemophilia: the need to support autonomy in children</i>	2016 Canada	4	Suggest to caregivers the implementation of motivational techniques to help the child or adolescent gradually advance to the autonomy of their care .	Qualitative study
Buckner TW, Witkop M, Guelcher C, Frey MJ, Hunter S, Peltier S et al. ¹⁵	<i>Management of US men, women, and children with hemophilia and methods and demographics of the Bridging Hemophilia B Experiences, Results and Opportunities into Solutions (B-HERO-S) study.</i>	2017 USA	3	To assess the needs of patients with hemophilia B, seeking to understand the psychosocial impact of hemophilia, including situations involving self-care .	Qualitative study
Jadhav U, Mukherjee K ¹⁶	<i>Assessment of healthcare measures, healthcare resource use, and cost of care among severe hemophilia A patients in Mumbai region of India</i>	2018 India	4	Associate the use of health resources and the cost of care through a scale that measures the patient's ability to maintain basic self-care in hemophiliac patients .	Qualitative study

Figure 2. Summary of publications included in the integrative review on self-care of hemophiliac children. Rio de Janeiro (RJ), Brazil, 2018.

After analyzing the articles, it was pointed out that it was possible to identify important points related to the self-care of the hemophilic child. The discussion was built in three major axes, generating a posteriori categorization of the exposed studies, as is foreseen in the methodology used in this work. The following categories were elucidated: “Motivation for self-management”; “Understanding and living with the diagnosis of hemophilia” and “Difficulty to perform self-care”. Articles were cited in more than one category, as evidenced by the scarcity of studies and the fact that the articles present information that adds to the various themes.

DISCUSSION

♦ Self-management motivation

Caregivers or parents of hemophilic schoolchildren should develop an integrative strategy that improves communication and motivation of children in managing their self-care. Through this strategy, healthy individual choices will be encouraged, as the child will understand his diagnosis and, therefore, will be more committed to his treatment.^{13, 14}

There are numerous benefits to patient empowerment through self-management from childhood, such as identifying bleeds, performing self-infusion, recognizing subtle body signs such as

bruising, bruising or tingling, and knowing when to request it. the help of adults or health professionals.¹³

In a recent study conducted at Hemocentro do Ceará, training was provided to people with hemophilia and their caregivers in order to encourage the practice of factor self-infusion, provide greater independence and reduce visits to treatment centers, obtaining a satisfactory result. with attitude.¹⁷

Such actions become indispensable for the autonomy of these individuals, referring to Dorothea Orem's Theory, where, through training, the patient makes his care more autonomous.⁴ This activity is corroborated by the American study,¹³ which evidenced that Haemophiliacs who provide self-management opportunities from childhood become experts in their disease and thus can keep it more easily under control, thus making it possible to minimize the impacts on the physical and psychological health of the haemophilic patient.¹⁸

It is understood that children need to develop strategies for risk management in everyday life, thus learning to judge and balance the possibilities and difficulties they may encounter, distinguishing them between negative and positive.^{13, 18} They are able, from the recognition of risks, to make more cautious decisions, even in the face of social

issues or impasses that may arise during the various stages of life.

It is influenced by the fear that children will be injured, the care provided to them, generating distressed parents / caregivers, controllers and overprotectors and, on the other hand, children lose several opportunities to find solutions to the daily challenges when facing their diagnosis. chronic disease.¹⁴

The Bridging Hemophilia B Experiences, Results and Opportunities into Solutions (B-HERO-S) study aimed to address some shortcomings related to the treatment of patients with hemophilia B.¹⁵ The study states that patients with severe hemophilia had more responsibility in their self-care, greater decision-making in face of the challenges of daily life and their treatment when compared to patients with moderate or mild hemophilia.

♦ Understanding and living with the diagnosis of hemophilia.

It is clarified by the World Hemophilia Federation,¹⁹ that patient education is essential for knowledge of their pathology, thus, they will be able to live and live with their health condition in a healthy way, becoming a citizen. productive and fully included in society. However, this knowledge should be extended to the caregiver family and, along with it, outline strategies that may assist in the management of chronic disease.

Some of the caregivers¹⁵ were found to use screening methods for bleeding episodes, such as electronic journals and spreadsheets (55%) or mobile phone applications (31%). This practice is advocated by other authors²⁰ who state that the use of these tools is the foundation for the organization of important data for the patient's health. With this strategy, access to data and identification of bleeding episodes become more practical. For example, it is possible to assess whether or not there was a need for factor administration or hospitalization.

Due to the systematic recording of these events, the responsibility for self-care is subjugated, generating a routine defended by the authors.¹⁵ It is clear that the diagnosis of chronic disease is a mixture of complex experiences that involve feelings linked to obstacles, often unpredictable. Therefore, there is an overload on caregivers, who are always concerned about the school and how certain actions can worsen their health.²¹ Thus becoming the record an important self-management document, which allows disclosure in consultations, presenting information more faithful of the episodes to health professionals involved in care, which could be forgotten if they had not been recorded.

It is believed that by establishing a habit, it is possible for the family to adapt to the new reality

and take care of the school in a holistic way, with a care routine that meets their basic needs, but at the same time, empowerment of their self-management, as indicated in Orem's Theory.⁴ The risk of accidents and bleeding is reduced by understanding the clinical condition, linked to the planning of daily life, allowing hemophilia not to become the central axis in patient's life and thus promote a higher quality of life as well as effective self-management.¹³

It is demonstrated that when there is no qualification of the health team and the family to treat the hemophilic child, it is exposed to situations defined as "attempts to get it right"¹⁶, that is, possible avoidable damages. The treatment is influenced by the difficulty in understanding the diagnosis by the caregivers and by the child, and this is noticeable in the case study¹² where damage to the health of the child was reported, given the poor performance of the care management. The theme will be better explained in the next category.

♦ Difficulty performing self-care

It is necessary to understand the social context in which these patients are inserted to understand the difficulties to perform self-care. It is noticed that the power of understanding and assimilation of the information received is different for people, because it is conditioned to their instructional and cognitive level.⁷ The studies that make up this category present completely different social contexts, which directly interfere with the ease or not access to health services and, consequently, maintenance Health care is provided in the US by the private sector, and in India it is organized by a government still under development.^{22, 23} It is noteworthy that in both countries there is difficulty in access to health and this directly influences the treatment of patients.

From this, it is pointed out by a study conducted in India, the reality of the country with regard to access to health and treatment for hemophilia, also mentioning how culture influences these issues. It was revealed in a survey conducted with 160 individuals, that 55% (n = 88) of the total number of respondents were negligent in self-care management, and several factors justifying this problem were listed.¹⁶

It is inferred that the difficulty in accessing the centers that treat hemophiliacs was the most relevant, not only because of the distance between their home and the care center, but also because of the cost related to travel and treatment. The authors considered treatment expenses over the 12-month period divided into direct (purchase of concentrates, hospitalization costs) and indirect (loss of salary due to crisis or recovery periods) expenses.¹⁶

The study also warns,¹⁶ that in India, there is a lack of specialized training for health professionals working with hemophiliacs, which generates a non-standardized treatment, causing problems to patients' lives that could be avoided if proper guidance was provided. It is regrettable, however, that the authors did not disclose the data, separating them by age group, being presented generally, which precludes a deeper analysis of the findings applied to the public chosen in this review.

In the American study,¹⁵ it is added that the data were not separated by age, however, the study shows a significant number of bleeding commonly caused by trauma (34%). It should be borne in mind that the occurrence of trauma can be prevented, if there is prior knowledge about its disease and the agents that cause trauma, in contrast, spontaneous bleeding cannot be predicted or prevented and represented 33 % of total number of respondents.

Another American study,¹² reported the case of a male child diagnosed with severe haemophilia A who had not been adequately treated until he was two years old and, as a result, had several episodes of hemarthrosis in both knees and decreased response to clotting factor replacement. The family was advised by the multidisciplinary team to perform more accurate tests to better understand the state of the disease, but the caregivers did not agree on the treatment indicated by the health team and postponed the tests, generating a new picture of the hemarthrosis and difficulty adhering. Adherence to the actions and behaviors of patients and caregivers related to access to consultations, lectures, support groups and the correct management of medications is related.²⁴

It is explained that the child¹² had to perform several surgeries in order to improve knee amplitude and, after the surgical intervention, some physiotherapy sessions where iatrogenesis occurred, leading to a fracture in the femur. Iatrogeny is defined as an unsatisfactory performance in care, which generates some "harm or harm to human well-being"²⁵ and can happen in any segment of the health area.

It is important to reflect that the decisions of parents or caregivers must be aimed at the well-being of the child, because their actions and choices are examples to be followed by their guardians. It may be difficult to maintain child care for their guardians to reflect on how that individual will plan their own care in the future; thus, each member of the hemophiliac care network, whether family or health professionals, needs to be in line with the assistance.

CONCLUSION

It is concluded that scientific publications related to current literature on how children with hemophilia A and B perform self-care were scarce. It identifies the need of the child to understand the disease and generate autonomy in the management of their self-care. Children need to understand that they are responsible for the consequences of their own behaviors, whether they are appropriate or not, and from this, make decisions about their health in a coherent and concise manner.

It is complemented, therefore, that Nursing plays an important role in the education of caregivers and children affected with chronic disease. In addition, the discourse needs to be strengthened through the publication of scientific evidence that corroborates its strategic actions aimed at the assisted public, generating motivation, understanding and, as a consequence, maintaining self-care.

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