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TAKING CARE OF HOSPITALIZED CHILD WITH WILSON'S DISEASE: EXPERIENCE REPORT[1]

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ABSTRACT
The hospitalization process can bring psychological and physical suffering to the child, especially when affected by a chronic and rare disease. This study aims to report the academic experience, emphasizing the importance of implementing the nursing process in assisting a child with Wilson's disease. This is a descriptive study of experience report type, developed at the Pediatric Clinic of a Teaching Hospital in the city of Joao Pessoa - PB. The results show that the academics were able to strategically develop nursing actions through the appropriation of information about the case and approach to the binomial, the conduction of actions for the nursing care and the realization of questioning on the care. This experience has allowed reflections on the care process, especially in cases of rare diseases in order to awakening in students the ability to “learn to learn” from practice.

Keywords: Nursing Process. Hospitalized Child. Chronic Disease. Hepatolenticular Degeneration. Nursing Education.

INTRODUCTION

The hospitalization process can bring children to destabilization of emotions and physical suffering, especially when they have a chronic and rare disease, as they remain restricted from their routine activities often for prolonged periods, being subjected to treatments and procedures that are difficult to be faced by a child as care(1,2).

The Ministry of Health (MoH) launched in 2014, the National Policy for Comprehensive Care to People with Rare Diseases of the Unified Health System (SUS), which aims to reduce morbidity and mortality rates of people with rare diseases (those that reach up to 65 people for each 100,000/inhabitants), in order to ensure universality, comprehensiveness and equity of health actions and services through a humanized care(3).

Among the group of rare genetic diseases, there is highlight for Wilson's Disease (WD), an autosomal recessive disorder caused by a mutation of the ATP7B gene, located on the long arm of chromosome 13 and characterized by reduced excretion of copper and, consequently, its accumulation in the body. This change occurs in the synthesis of the protein responsible for copper entrainment into the extracellular medium, whose main source of elimination takes place through the bile. It has a prevalence of 1:30,000-100,000; however, in some countries, such as Brazil, this prevalence is not yet established(4-6).

This defect in copper transport leads to the intracellular accumulation, exceeding the physiological level of storage, causing damage to the liver, to the central nervous system, in kidney and cornea, in addition to cardiac abnormalities. The classic triad of the disease consists of liver, neurological and ophthalmologic changes(4,5). In children, it can appear after three years of age; however, the diagnosis is complex in this age group and may be associated with any finding of change in liver function or chronic disease, such as hemolytic anemia and acute liver failure(7).

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[1] It was extracted from the research projec ttitled: Nursing Care Systematization for children with chronic disease. This project is linked to the Study and Research Group on Child and Adolescent Health and to the Study and Research Group “Grounds of Nursing Care”; both groups are attached to the Graduate Nursing Program of the Federal Universityof Paraíba.

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Research conducted in southern Brazil has identified 14 different mutations in the WD, differentiating treatment(6), which is based on chelating drugs of copper and zinc salts, used in monotherapy or combined therapy. It has proved effective, without which the disease becomes lethal. Therefore, early diagnosis has a significant impact on quality of life of these patients(4,8,9).

In this sense, nursing care becomes crucial, as it seeks the promotion and restoration of the individual's quality of life in their biopsychosocial dimensions, causing the increase in life expectancy(10). Recognizing the critical thinking as a differential tool in care actions is fundamental to the care process. By considering that the nursing practice requires skills, and that these need to be developed through the implementation of the nursing process, it is necessary to insert this process in academic nursing education as a way to awaken the critical and reflective sense about the care.

In the practical and theoretical activities of the discipline Nursing in the Health Care of Children and Adolescents II (DESCA-II) of the undergraduate nursing degree from a public institution, the student is inserted in care practices to deal with the encountered difficulties, meeting the demands of hospitalized children and adolescents and their families. In this perspective, the question is: is it possible to implement the nursing process in the academic activities of care to the binomial child/family with rare disease as a learning tool, whereas this process helps in decision-making, in the acquisition of autonomy and responsibility, besides being a methodological tool that guides the professional practice regulated by Resolution No. 358/2009 of the Federal Council of Nursing (COFEN)?

The literature has a gap regarding the description of nursing care to children with WD. Thus, authors sought to associate this gap to the implementation of the nursing process as a tool in child care. Therefore, this study aims to report the academic experience in the care of a child with Wilson's disease and their family, through the implementation of the nursing process as a learning strategy.

METHODOLOGY

This is a descriptive study, of experience report type, resulting from academic experience, in which care actions were effected to an eight–year-old male student with WD, and to his family. It should be noted that this boy was chosen by considering the rarity of the case in children and the complexity of the case in this age group.

The care actions took place in the Pediatric Clinic of the Lauro Wanderley University Hospital (HULW), located in Joao Pessoa-PB, during theoretical and practical activities carried out in March 2014, under the guidance and monitoring of teachers and monitors of the DESCA-II, with the collaboration of nurses of the clinic.

The HULW is considered a reference for the treatment of chronic and rare diseases, and it is a field of utmost importance for the process of training students in the health and other similar areas, since it contributes to building critical and reflective sense, and the development a broad view of the care process and the work process.

For the preparation of the experience report, authors used the discipline reflective diary notes and structured observations with the aid of a data collection instrument and patient's records (admission history, daily activities of the nursing team and annotations of the health team). No personal data were used, only those of pathophysiological relevance to the description of the experiment. It is also noteworthy that the legal guardian has signed an Informed Consent Form allowing the dissemination of the experience.

The dynamics used for data analysis and description of the experience is based on the phases of the nursing process, as regulated by Resolution No. 358/2009 of COFEN: data collection, nursing diagnosis, nursing planning, nursing implementation and evaluation, as well as the assumptions of the theory of basic human needs of Horta, seeking to develop in the student process of action/reflection/action in the context of healthcare reality of the health service in a hospital setting, by basing care actions in the relevant literature about the theme.

It is noteworthy that the report met the ethical principles of Resolution 466/12 of the

National Health Council (CNS), which deals with the Guidelines and Norms Regulating Research Involving Human Beings, and the Code of Ethics of Nursing Professionals, Resolution 311/2007. The report is resulting from a research project developed with children and adolescents with chronic disease approved by the Research Ethics Committee of the Hospital under the Protocol 222/09, CAAE-0052.0.126.000-09, which is linked to the Research Group on Child and Adolescent Health and to the Research Group Based on Nursing Care, both linked to the Graduate Nursing Program of the Federal University of Paraiba.

RESULTS ADN DISCUSSION

When entering the student into professional practice in order to make them think about their care practice, teachers need to adopt strategies that stimulate curiosity and ensure the involvement between theory and practice. Thus, the adoption of the nursing process as a strategy to guide the care actions contributes to building skills, critical thinking, autonomy and interpersonal relationships for the training of the academic, who is encouraged to implement strategies from a systematized practice and based on scientific knowledge(1).

Given the context of experience, three moments were highlighted during the service: Appropriation of information about the case and approach the binomial; Actions for nursing care; and Reflections on care, in which the phases of the nursing process are inserted. It is noted that the implementation phase (interventions) is described in the second part of the experience.

Appropriation of information about the case and approach the binomial

The approximation to the binomial is inherent to the first phase of the nursing process (data collection), a condition that brings us questions: what should we ask? How should we interact? What made them require hospitalization? The search for answers was given in consultations, in the medical record and subsequently in the literature. Therefore, access to information and communication technologies was crucial in the practical field; multifunctional mobile phones were used for quick consultation of texts in order to contribute to learning and care, which helped in the appropriation of information on the clinical picture.

Understanding the child's clinical picture facilitated the data collection process, and the establishment of relations with the binomial, which are important conditions for the establishment of the bond and trust, because, at any moment, many questions about the disease and the likely treatment were imposed for by the binomial, and it was necessary to create strategies to convey information and enhance understanding of the child and the companion on the pathology, treatment or consequences of its abandonment.

Although the literature presents the neurological signs and the Kayser-Fleischer Ring (metallic impregnation in the cornea) as the most suggestive changes, such signs were not detected on physical examination, nor on the results of tests. In most children and adolescents, these signals are absent in the early development of the disease(5).

The WD is a rare disease and its diagnosis is not simple(6). As a result, the population has no information about it and, therefore, the professional should clarify the doubts, relieving anxiety and favoring the formation of the bond, allowing a more accurate data collection with more accurate nursing diagnoses.

In this context, listening and dialogue, provided in the data collection phase, with regard to the child's health process, enables creating a link between the nursing staff and the binomial, so that this condition allows a care that softens the physical and emotional discomfort, by meeting the needs not only of the child but of their family, too(11).

After the approach and the establishment of the first contact, the trust relationship between students and the binomial was established. In the child, researchers observed feelings of joy and safety, which were expressed in gestures of affection and jokes, which allowed greater acceptance and effective participation in the realization of interventions performed. As for the mother, researchers observed a helpful and friendly companion, involved in the care of the child and cooperating in data collection.

Actions for nursing care

Identifying problems, developing diagnostics and thinking what to do, that is, preparing the planning of nursing care triggered many feelings, but with a proper collection, the care focused on the learning needs for health and therapy, (there was lack of knowledge about the disease and treatment), with prescriptions, mostly aimed at orientation. Thus, the care plan was directed to Health Education (HE), as it allows a critical reflection of reality, enhancing our ability to provide care and promote health. In addition, HE actions are more constructive, reflective and dynamic(12).

From this perspective, it is emphasized the importance of guiding the family from the perspective of HE, since by providing education, nurses promote health. Therefore, it is necessary that the professional listens the patient attentively and warmly in order to answer questions, relieve pain and anxiety, by explaining processes and procedures. These attitudes will contribute positively in the child's quality of life and will provide a more effective therapy adherence by the family. Moreover, the attitudes of the future professional will be enriched.

In pediatrics, the most common clinical manifestations of WD are the liver changes. The disease may be in acute, chronic or fulminant forms. In the acute form, the patient has elevated levels of aminotransferases, jaundice and hepatomegaly. In the chronic form, there are signs of portal hypertension and hepatosplenomegaly, as well as elevated liver enzymes. In turn, the fulminant form is considered when the values of serum, urine and liver copper are at high levels, as well as bilirubin(4,13). It is worth noting that there are cases of patients who have the disease in its acute form, but are endowed with absence of signs and symptoms or even, there is presence of suggestive symptoms of very mild liver disease(4).

Such understanding was necessary because the implementation of nursing interventions was focused to clarify the binomial about these findings, with focused guidance to the psychobiological and psychosocial care, by emphasizing the regression in the size of organs after starting the medication, which aimed at emotional stabilization of the binomial.

In this perspective, the inclusion of the family in the care plan was essential, considering its individuality and seeking to realize the family member and the child as one. This approach provides stability and strength to the family and the binomial during periods of hospitalization(14). Adherence to interventions became more visible as the child realized that the guidelines had to do with the symptoms he had, namely, liver and spleen changes (hepatosplenomegaly).

In addition to the guidelines, it was necessary to perform procedures, such as administering medication, monitoring side effects and guiding the binomial on the disease. It was a difficult time because the medications for WD are not part of the service routine and thus required the inclusion of the child in the State register for subsequent acquisition. Ordinance No. 848/2011 states that, for dispensation of drugs required under the Clinical Protocol and Therapeutic Guidelines of Wilson's Disease, the patient must meet the criteria described in the protocol(15).

According to the protocol of therapeutic guidelines for WD, the D-Penicillamine, a copper-chelating drug, acts by removing and detoxifying intra and extracellular copper. There is a table with three dimensions of evaluation: signs and symptoms, laboratory tests and analysis of mutations, in which the patient whose sum is four points or more is included in the group of patients who should receive treatment for WD(15). The patient had suggestive neuropsychiatric symptoms (two points), negative Coombs test (one point) and serum ceruloplasmin less than 10 mg/dl (two points), totaling five points.

Importantly, in the management process of D-Penicillamine, the patient may have several adverse effects, which can be fatal. So, throughout the administration of the drug, this must remain under observation in hospital(9,15). For this reason, the patient or the guardian must sign the Informed and Responsibility Form on the use of Penicillamine and Trientine, described in the above ordinance.

It should be noted that, prior to administering the drug, the child and the family were told about the procedure, the treatment time and the unwanted reactions that the medication could cause. Still, they were guided as the team's readiness to explain questions.

It is worth mentioning that the child did not show signs and symptoms of side effects while in the hospital environment, which contributed potentially to his discharge and the regression of clinical (liver and spleen) signs. When the binomial was informed about the positive response to medication, both showed great happiness. This was one of the most gratifying moments for scholars throughout the care.

Other interventions were focused on games, a method that alleviates the suffering of hospitalization and improves the environment, making it more enjoyable. Prior to and at the end of the procedures, it was possible to play, which gave the child a notorious happiness. The Therapeutic Play (TP) technique alleviates the suffering of the child, making care less traumatic(16), and is a technique recommended by COFEN in pediatric care, according to Resolution 295/2004.

Reflections on the care

The evaluation of nursing care was carried out in order to provide improvements to care and contribute to the learning process of the academics regarding the systematization of care to the binomial. From the practice, it was possible to develop a critical reflection on the qualified listening, by offering individualized and humanized care, on knowledge and autonomy acquired during the implementation of the nursing process. In this sense, the reflections on the care were held in a discussion wheel, in which students exposed the situations experienced, by promoting critical and reflective dialogue about each strategy implemented, culminating in the redefinition of their practice. The implementation of interventions was enriching because students were refuted before the doubts that have arisen, which motivated them to search for new knowledge.

The assistance provided was assessed as positive by all actors involved in the care process (teachers/students/binomial), since the care to the binomial needs to consider the patient and their family as one, requiring comprehensive care.

Therefore, enhancing and strengthening the actors involved in the care process by recognizing their role as health promoters(17) arouses feelings of shared responsibility with care. It should also be highlighted the importance of the professional in supporting the family so that they can understand the whole process and continue taking care of the child.

FINAL CONSIDERATIONS

The nurse has an essential role in health prevention, promotion, recovery and treatment of children, which requires the relationship with the binomial. So that those health actions are achieved, it is necessary that the professional has a wide view of care and positions themselves as an active character in the care process for hospitalized children. Therefore, the sense of care exceeds the technical doing and the mere execution of routines, tasks and protocols; it needs to be conducted in a comprehensive manner(16).

The findings of this study seek to report the experience with an assisted child in local reality. So it does not allow generalization, given the individuality of the nursing care to the hospitalized child. However, the experience report aims to collaborate in the construction of knowledge about the process of taking care of children affected by WD and contribute to the training process of nurses, encouraging them to meet the binomial in their biopsychosocial aspects in a critical and reflective manner.

CUIDANDO DA CRIANÇA HOSPITALIZADA COM DOENÇA DE WILSON: RELATO DE EXPERIÊNCIA

RESUMO

O processo de hospitalização pode trazer à criança sofrimento psicológico e físico, principalmente quando acometida por uma doença crônica e rara. Este trabalho tem como objetivo relatar a experiência acadêmica, enfatizando a importância da implementação do processo de enfermagem na assistência a uma criança com Doença de Wilson. Trata-se de um estudo descritivo, tipo relato de experiência, desenvolvido na Clínica Pediátrica de um Hospital Escola do município de João Pessoa-PB. Os resultados demonstram que foi possível aos acadêmicos a elaboração de maneira estratégica das ações de enfermagem por meio da apropriação de informações sobre o caso e aproximação ao binômio; da efetivação de ações para o cuidado de enfermagem e da realização de ponderações sobre a assistência. Essa experiência permitiu reflexões a respeito do processo de cuidar, principalmente em casos de doenças raras, no intuito de despertar no discente a capacidade de “aprender a aprender” a partir da práxis.

Keywords: Nursing Process. Hospitalized Child. Chronic Disease. Hepatolenticular Degeneration. Nursing Education.

CUIDANDO AL NIÑO HOSPITALIZADO CON ENFERMEDAD DE WILSON: RELATO DE EXPERIENCIA

RESUMEN

El proceso de hospitalización puede llevar al niño sufrimiento psicológico y físico, sobre todo cuando acometido por una enfermedad crónica y rara. Este estudio tiene como objetivo informar sobre la experiencia académica, haciendo hincapié en la importancia de la aplicación del proceso de enfermería en el cuidado a un niño con la Enfermedad de Wilson. Se trata de un estudio descriptivo tipo relato de experiencia, desarrollado en la Clínica Pediátrica de un Hospital Escuela, de la ciudad de João Pessoa – PB, Brasil. Los resultados muestran que fue posible para los académicos la elaboración de forma estratégica de las acciones de enfermería a través de la apropiación de informaciones sobre el caso y aproximación al binomio; de la efectuación de acciones la atención de enfermería y la realización de consideraciones sobre la atención. Esta experiencia permitió reflexiones sobre el proceso de cuidar, sobre todo en los casos de enfermedades raras, con el objetivo de despertar en el estudiante la capacidad de aprender a aprender a partir de la praxis.

Palabras clave: Procesos de Enfermería. Niño Hospitalizado. Enfermedad Crónica. Degeneración Hepatolenticular. Educación en Enfermería.

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