Health literacy: educational brochure for pediatric patients with neurofibromatosis type 1

Letramento em saúde: fôlder educativo para pacientes pediátricos com neurofibromatose tipo1

Alfabetización en salud: folleto educativo para pacientes pediátricos con neurofibromatosis tipo 1

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Abstract

Objective: Develop a folder for caregivers of pediatric patients newly diagnosed with neurofibromatosis type 1 and for adolescents and young adults living with the disease.

Methods: Non-experimental descriptive study, using empirical action research. The process of creating the material occurred in three stages: identification of the problem; team discussion; and compilation of knowledge and production of the folder.

Results: The folder prepared presented basic information about the disease, with concepts and figures. It also warned the reader about searching for information on the internet, which may be inadequate, non-specific or without scientific evidence.

Conclusion: The use of health literacy as an education tool is reflected in decision-making about health and necessary medical care. The knowledge obtained from reading materials like this increases autonomy in relation to treatment and health decisions, as more reliable knowledge about the disease is acquired.

Keywords

Neurofibromatosis 1; Health literacy; Rare diseases; Patient education handout; Pediatrics

Resumo

Objetivo: Desenvolver um fôlder para cuidadores de pacientes pediátricos recém-diagnosticados com neurofibromatose tipo 1 e para adolescentes e adultos jovens que convivem com a doença.

Métodos: Estudo descritivo não experimental, por meio de pesquisa-ação empírica. O processo de criação do material ocorreu em três etapas: identificação do problema, discussão em equipe e compilação do conhecimento com produção do fôlder.

Resultados: O fôlder elaborado apresentou informações básicas sobre a doença, com conceitos e figuras. Também alertou o leitor sobre a busca por informações na internet, as quais podem ser inadequadas, inespecíficas ou sem evidência científica.

Conclusão: A utilização do letramento em saúde como ferramenta de educação se reflete na tomada de decisão sobre saúde e cuidados médicos necessários. O conhecimento obtido a partir da leitura de materiais como este aumenta a autonomia diante do tratamento e das decisões em saúde, visto que se adquire conhecimento mais fidedigno sobre a doença.

Descritores

Neurofibromatose 1; Letramento em saúde; Doenças raras; Prospecto para educação de pacientes; Pediatria

Resumen

Objetivo: Desarrollar una carpeta para cuidadores de pacientes pediátricos con diagnóstico reciente de neurofibromatosis tipo 1 y para adolescentes y adultos jóvenes que viven con la enfermedad.

Métodos: Estudio descriptivo no experimental, mediante investigación acción empírica. El proceso de creación del material se desarrolló en tres etapas: identificación del problema; discusión en equipo; y recopilación de conocimientos y producción de la carpeta.

Resultados: La carpeta elaborada presentó información básica sobre la enfermedad, con conceptos y cifras. También advierte al lector sobre la búsqueda de información en Internet, que puede ser inadecuada, inespecífica o sin evidencia científica.

Conclusión: El uso de la alfabetización en salud como herramienta educativa se refleja en la toma de decisiones sobre la salud y la atención médica necesaria. El conocimiento obtenido a partir de la lectura de materiales como este aumenta la autonomía en relación con el tratamiento y las decisiones de salud, ya que se adquieren conocimientos más fiables sobre la enfermedad.

Descriptores

Neurofibromatosis 1; Alfabetización en salud; Enfermedades raras; Folleto informativo para pacientes; Pediatría

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Introduction

Neurofibromatosis type 1 (NF1) designates a genetic disease within the scope of systemic phakomatoses, with an autosomal dominant inheritance pattern. Phenotypically, the disease is expressed by multiple *café-au-lait* spots, various neurofibromas, ephelides, bone dysplasia, among other changes, with varying complexity, including in individuals from the same family.[1,2]

Data from the United Kingdom Neurofibromatosis Association Clinical Advisory Board estimate an incidence of approximately 1:3,000 cases, with no statistically significant difference between certain populations and gender.[2] As it is a genetic disease with an autosomal dominant inheritance pattern, each person affected by NF1 has an estimated 50% chance of transmission, with its phenotypic manifestation being close to 100%. However, there is the possibility that the disease is generated by a de novo pathogenic genetic variant, i.e., the pathogenic variant was not inherited from the parents and appeared for the first time in that individual.

Diagnosis is predominantly clinical, regardless of its genetic origin, and is based on phenotypic criteria of the disease, although genetic tests are important in identifying pathogenic variants (previously called mutations) specific to the disease.[1,2] Thus, an in-depth anamnesis with a well-defined genogram, coupled with an accurate physical examination, tends to be sufficient for NF1 diagnosis.

However, after diagnosis, routine examinations are essential to assess the extent of the disease and its monitoring, in order to identify and intervene early if complications arise. Treating this condition is still a challenge,[1-3] since its cure has not yet been found. However, it is possible to treat each of the complications of the disease individually, according to its extent and the individuality of each patient.

Disease diagnosis is, in most cases, cold and lonely, as patients who live with NF1 and their family experience a mix of emotions and feelings generated either by fear of an uncertain prognosis or by guilt. An interprofessional approach with these patients is essential not only for monitoring their physical health, but also for their emotional health. With diagnosis, many doubts and uncertainties are present, both due to the different signs and symptoms of the disease as well as its uncertain prognosis and lack of knowledge about how the disease is inherited, which is why a feeling of guilt arises in parents. Facilitated access to information on the internet contrasts with the collection of true and/or inadequate information about NF1, which can increase fear of the disease.

Nowadays, the term health literacy (HL) has been growing and taking up space in world literature. The World Health Organization (WHO)[4] considers that HL represents the knowledge and personal skills that individuals accumulate through their personal and collective experiences, which enable people to understand, assess and use information and services in ways that promote and maintain health and well-being for themselves and those around them. With this, HL has become fundamental for decision-making, based on inclusive and equitable access and being part of health promotion.[4]

In the area of nursing, the NANDA-I Nursing Diagnosis (ND) "Readiness for enhanced health literacy" is defined as a pattern of use and development of a set of skills and competencies (literacy, knowledge, motivation, culture and language) to find, understand, assess and use health concepts and information for daily decision making, health promotion and maintenance, health risk reduction and overall quality of life improvement, which can be improved.[5]

Nurses, as nursing team leaders, must recognize patients and their support network as key points of attention when planning the systematization of nursing care so that, with the multidisciplinary team, they can outline educational strategies. Educational actions not only help improve adherence to proposed treatments, but also bring a sense of greater autonomy to patients and all those included in the educational process.

To date, studies on HL of patients with NF1 or on the topic with their parents or caregivers are not found in national or international scientific article repositories. For adequate monitoring of NF1 to be effective and assertive of patients' individuality, it is essential to train them, as well as their families, with the main themes and concepts of the disease, allowing their participation in the entire health-disease process that involves them. This study aimed to develop a folder for caregivers of pediatric patients newly diagnosed with NF1 and for adolescents and young adults living with the disease.

Methods

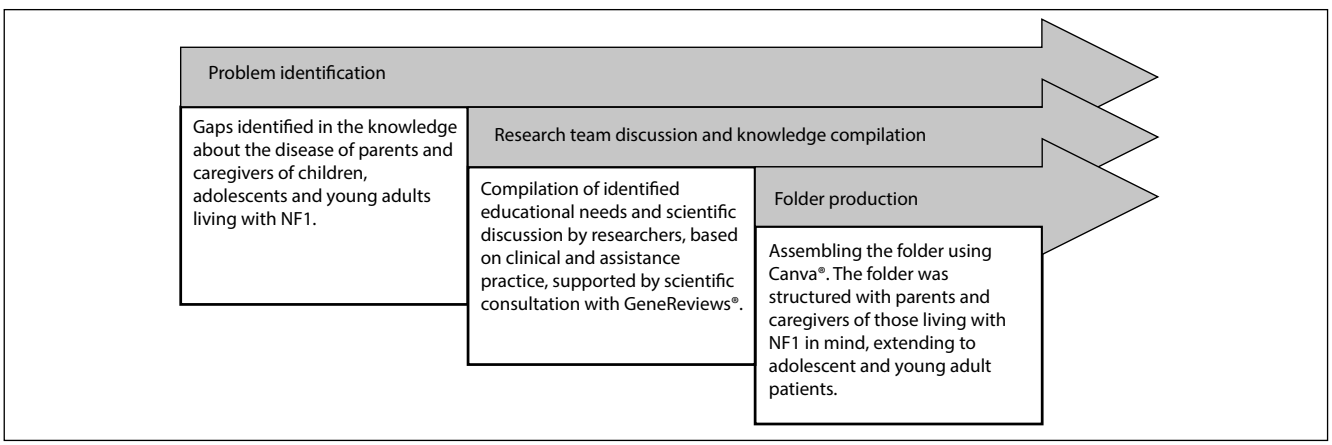
This is a non-experimental, descriptive study, with a view to developing an educational folder. The methodology used to create the educational material was empirical action research, as it accumulates data from experiences to develop foundations. It is a study model widely used in the educational field, as it enables information and knowledge production, whose main objective is knowledge construction through collaborative self-reflection between groups seeking to rationalize social and educational practices.[6-8]

Through action research, it is possible to find an investigation approach that mixes academic research with practice to promote social transformations, generating knowledge applicable to the study question. In this sense, action research is defended from a theoretical-methodological perspective that allows it to overcome the logic of technical rationality through knowledge production.[8]

Therefore, this study was developed through discussions between a team of researchers from the Child Health Care research line at a federal university in southern Brazil; such discussions discussed the gaps found in knowledge about NF1 of parents and caregivers of children, adolescents and young adults living with the disease, identified during data collection for a matrix study entitled "Quality of Life and Health Literacy of Patients with Neurofibromatosis Type 1 and the Impact of the Disease on the Family".

In the construction of this manuscript, the Equator Network SQUIRE 2.0 guideline was used, which offers a model to describe new knowledge on how to improve and improve health actions. SQUIRE 2.0 describes work done at a system level to improve the quality, safety and value of healthcare, among others.[9] The material was developed between August and October 2023. A total of 18 parents or guardians of pediatric patients living with NF1 participated. Recruitment took place through a call on social networks, such as Facebook® and Instagram®, in which an invitation was made to report the signs and symptoms they knew about the disease and present their greatest doubts regarding NF1. The doubts and questions presented were summarized and answered by consulting GeneReviews®.[2] GeneReviews® is an electronic platform that provides review articles that describe specific genetic diseases, which are standardized, peer-reviewed and constantly updated. Figure 1 represents the process of preparing the material.

It is important to highlight that there was no content validity stage for the material, as its creation was the result of the needs identified in the matrix study described. Since its completion, the folder has been distributed at scientific events to healthcare professionals who assist this population as well as to parents and guardians of patients with NF1 in outpatient consultations. The material aimed to widely disseminate basic information about NF1, helping to better understand patients and their families, despite both the genetic aspects of the disease and its heredity and clinical signs. It was extracted from the results of a non-profit project, financed by the authors themselves. The project that led to educational folder development was ap-



NF1 - neurofibromatosis type 1

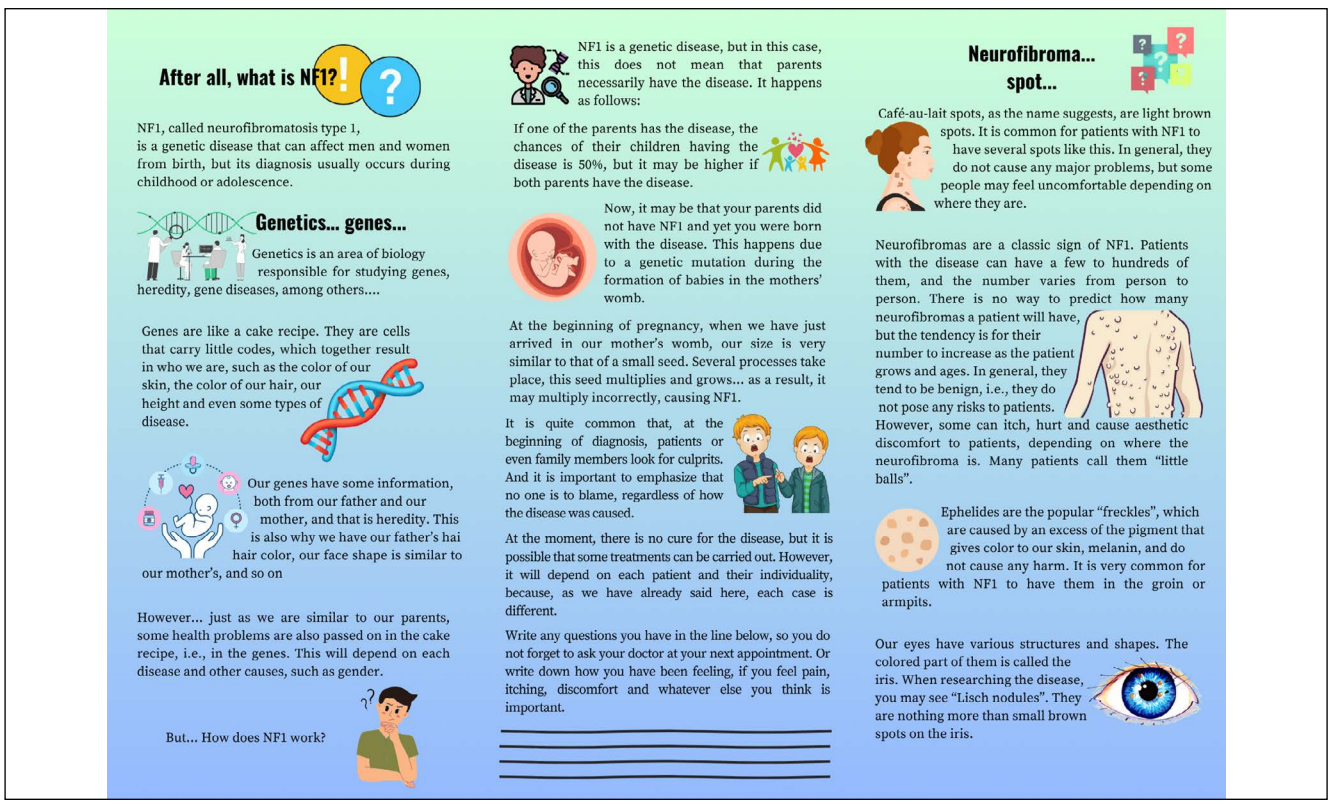
Figure 1. Stages of the educational material creation process

proved by the Research Ethics Committee of the proposing institution through Opinion 5800810 (CAAE 65440322.0.0000.5335).

Results

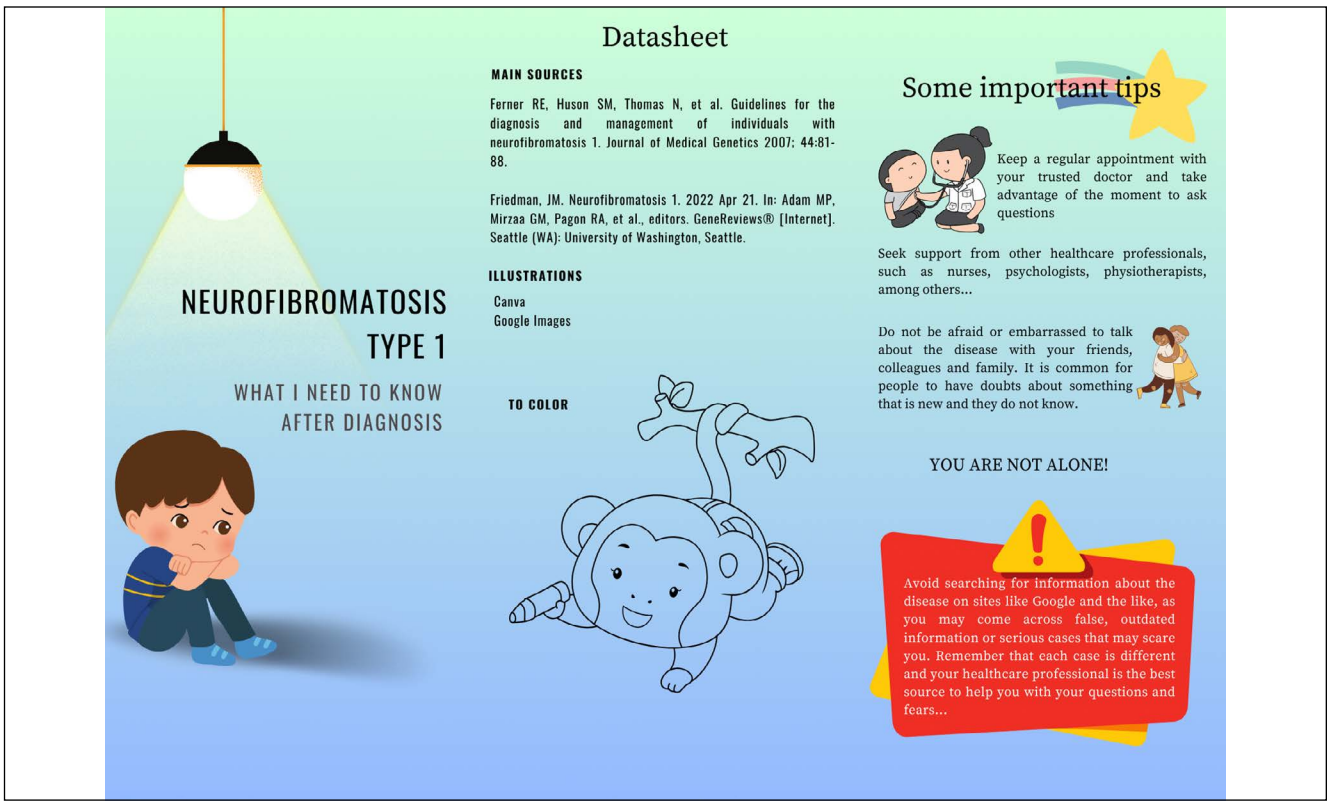
The product developed had dimensions of 29.7x21 cm, was made in the three-fold style and developed with predominantly pastel colors. Two details caught attention on the obverse. The first was to place a drawing for children to color, with the aim of bringing younger patients closer to the material, arousing not only their curiosity, but also making them feel part of their health-disease process. The second was an alert to be careful when searching for information about NF1 on different websites, something that could be common for older children, teenagers and even their families and caregivers. This was an important warning, as they may come across inadequate information about disease course or prognosis as well as non-specific information or without scientific evidence, which could scare them. Reinforcing that each case was different from the other is important in the recent diagnosis phase, as it prevents patients and their support network from becoming frightened or blaming themselves due to the information accessed, bringing patients closer to multidisciplinary monitoring. Figure 2 shows the obverse of the material.

On the back of the material, it was possible to find general information about the disease and its genetic inheritance. Many patients experience the anguish of being the first case in the family, afraid of the uncertain and unknown disease prognosis. Moreover, there was also the feeling of guilt that many parents experienced when having a child with NF1. Making them aware of how the disease was transmitted between family members and how new transmissions occurred is important so as not to affect the relationship between family members at the time of investigation and diagnosis. Furthermore, it was important to provide guidance on the terms and main findings of the disease, which were neurofibromas and *café-au-lait* spots (main findings of NF1). Figure shows the back of the folder.



Type 1 - What I Need to Know After Diagnosis

Figure 2. Obverse of the folder entitled Neurofibromatosis



Type 1 - What I Need to Know After Diagnosis

Figure 3. Back of the folder entitled Neurofibromatosis

Discussion

Educational materials for patients with chronic illnesses are essential. They have the role not only of education itself, but also of bringing patients, even pediatric ones, and their guardians closer to their illness and the necessary care. There is a lack of studies in the literature on HL in people with NF1 and their caregivers. Materials such as the folder presented have also been designed in other configurations and formats by Brazilian researchers, such as the booklet *As Manchinhas da Mariana*,[10] of the Minas Gerais Association of Support for Patients with Neurofibromatosis (Amanf - *Associação Mineira de Apoio aos Portadores com Neurofibromatoses*), in partnership with the Reference Center for Neurofibromatosis of the *Federal University of Minas Gerais*, and the guide *Tudo sobre Neurofibromatose Tipo 1*,[11] from Beaba – Civil Society Organization of Public Interest. Both materials aim to educate patients, family members and caregivers about the main aspects of the disease, with their nomenclatures, illustrations and meanings.

An international multicenter study described the development of educational materials for parents and caregivers of children with cancer, health education aiming to reduce acceptance barriers in the inclusion of children in palliative care.[12] The research highlighted the potential to improve access and acceptance of palliative care in families of children with cancer, reinforcing the importance of multidisciplinary studies for developing educational materials for patients and families. The authors state that the materials generated serve as a model for developing similar resources on other health topics to improve HL, especially in low- and middle-income countries.[12] We pursue this same approach in NF1.

There are, in the scientific literature, reports on the development of other educational materials for HL of children and adolescents with chronic illness. One of them was a Brazilian study,[13] which aimed to build and validate educational videos as part of the LISA Down Program – Literacy and Innovation in Health for Adolescents with Down Syndrome. This is material approved by both expert judges and its target audience, being used as an educational strategy in routines developed by institutions that care for pediatric patients with Down syndrome.

An integrative review study on HL for caregivers and people with rare diseases[14] shows the scarcity of the topic in the context of these diseases, especially in the Brazilian reality. This same conclusion is also exposed by the LISA Down research team.[13] Silva *et al*.[13] and Teixeira *et al*.[14] reflect on the scarcity of research on HL with the pediatric public. Inadequate HL can cause harm in the population's health context,[15] making it necessary to carry out more in-depth studies of HL in the pediatric population with NF1, their parents and caregivers.

The multidisciplinary work to improve HL is reinforced both by national researchers[13-15] and by an international multicenter study.[12] Creating educational materials in the form of folders is intended to reach a larger portion of the population, as they are instruments that can be used by healthcare professionals in clinical and care services.[16] Furthermore, based on the NANDA-I ND "Readiness for enhanced health literacy",[5] nurses can use educational materials as part of nursing care, through health education, both in hospital nursing care and in outpatient nursing consultations or in Primary Health Care.

The limitation of this study was that content validity was not carried out with the target population or with experts. Furthermore, studies on HL with pediatric and adult patients living with NF1 were absent. Studies on HL with the target population would present a greater scope on the topic, guiding researchers and healthcare professionals to manage health education in these patients. They would also help direct nursing care and develop educational products aimed at the specific needs of the population in question. Although the material content is simple and cannot be used as a protocol or therapeutic guideline, it can complement educational actions aimed at parents and caregivers of patients with NF1 as well as to patients with more advanced age and cognitive development, such as adolescents and young adults. On the other hand, simplicity makes the folder easier to accept and read.

Conclusion

Using HL as a tool to empower individuals to obtain, process and understand information and basic health services will be reflected in decision-making, both about their health and the medical care they need or will need. Thus, HL, quality of life and the impact of the disease on the family are key factors in improving these patients' adherence to follow-up, avoiding possible complications. The development of teaching material aimed at specific populations, as in the case of NF1, helps to fill this gap. Studies are needed to deepen knowledge about HL of patients and their guardians in the context of NF1 as well as strategies to better educate them. The material presented in this article has not undergone a validity process. However, creating a folder on NF1 helps to provide information about the disease to patients and their families in a fun way. The knowledge obtained from reading materials like this increases patient autonomy regarding treatment and health decisions, as they acquire more reliable knowledge about the disease. Therefore, studies on quality of life and the impact of the disease on the family are necessary so that we can make progress on the topic of NF1. This will not only allow the creation of more effective public policies aimed at this population, but also a better inclusion of these patients in the various health services.

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Collaborations

Souza LP and Zen PR contributed to project design, data analysis and interpretation, article writing, critical review of relevant intellectual content and approval of the final version to be published.

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