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COPING STRATEGIES IN THE EXPERIENCE OF MOTHERHOOD AGAINST EPIDERMOLYSIS BULLOSA

ESTRATÉGIAS DE ENFRENTAMENTO NA VIVÊNCIA DA MATERNIDADE FRENTE À EPIDERMÓLISE BOLHOSA

ESTRATEGIAS DE ENFRENTAMIENTO EN LA VIVENCIA DE LA MATERNIDAD FRENTE A LA EPIDERMÓLISIS BULLOSA

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Objective: to know the coping strategies used by the mother of an infant with Epidermolysis Bullosa. Method: a single case study with a qualitative approach conducted with the mother of a six-month-old child with clinical diagnosis of Epidermolysis Bullosa. Data collection occurred between February and March 2020 at the participant's home, through in-depth interviews and observation. The narratives were submitted to thematic content analysis. Results: thematic categories emerged, being "(Re)meaning motherhood after the diagnosis of Epidermolysis Bullosa", which addresses the moments of discovery of the rare disease, as well as the use of the internet as a resource for coping, that presents the movement to acquire theoretical knowledge to enable maternal care performed to the child. Conclusion: the study allowed the understanding of the coping strategies used in the experience of motherhood before the birth of the child diagnosed with Epidermolysis Bullosa.

Descriptors: Rare Diseases. Epidermolysis Bullosa. Family Power. Child Care. Pediatric Nursing

Objetivo: conhecer as estratégias de enfrentamento utilizadas pela mãe de uma lactente com Epidermólise Bolhosa. Método: estudo de caso único com abordagem qualitativa realizado com a mãe de uma criança com seis meses de idade e diagnóstico clínico de Epidermólise Bolhosa. A coleta de dados ocorreu entre fevereiro e março de 2020 no domicílio da participante, por meio de entrevista em profundidade e observação. As narrativas foram submetidas à análise de conteúdo do tipo temática. Resultados: emergiram categorias temáticas, sendo elas "O (res) significar da maternidade após o diagnóstico de Epidermólise Bolhosa", que aborda os momentos de descoberta da doença rara, bem como a utilização da internet como recurso para o enfrentamento, que apresenta o movimento para aquisição

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de conhecimentos teóricos para possibilitar o cuidado materno realizados à criança. Conclusão: o estudo permitiu apreender as estratégias de enfrentamento utilizadas na vivência da maternidade diante do nascimento da criança com diagnóstico de Epidermólise Bolhosa.

Descritores: Doenças Raras. Epidermólise Bolhosa. Poder Familiar. Cuidado da Criança. Enfermagem Pediátrica

Objetivo: conocer las estrategias de afrontamiento utilizadas por la madre de una lactante con Epidermólisis Bullosa. Método: estudio de caso único con enfoque cualitativo realizado con la madre de una niña de seis meses de edad y diagnóstico clínico de Epidermólisis Bullosa. La recogida de datos tuvo lugar entre febrero y marzo de 2020 en el domicilio de la participante, a través de una entrevista en profundidad y observación. Las narrativas fueron sometidas al análisis de contenido del tipo temático. Resultados: surgieron categorías temáticas, siendo ellas "O (res)significar de la maternidad después del diagnóstico de Epidermólisis Ampollar", que aborda los momentos de descubrimiento de la enfermedad rara, así como la utilización de internet como recurso para el enfrentamiento, que presenta el movimiento para adquirir conocimientos teóricos para posibilitar el cuidado materno realizado a la niña. Conclusión: el estudio permitió aprehender las estrategias de enfrentamiento utilizadas en la vivencia de la maternidad ante el nacimiento del niño con diagnóstico de Epidermólisis Ampollar.

Descriptores: Enfermedades Raras. Epidermólisis Espinosa. Poder Familiar. Cuidado del Niño. Enfermería Pediátrica

Introduction

Epidermolysis bullosa (EB) is characterized as a rare, non-contagious disease, in which the main characteristic is the fragility of the skin and mucous membranes of the body, which manifests itself mainly with the appearance of blisters and lesions in the body[1-2]. It is noteworthy that there are currently no epidemiological data on its distribution in Brazil, mainly due to difficulties in the location and distribution of these cases[3-4].

In the hereditary form of EB, mutations occur in structural proteins that anchor the skin, representing a severe clinical picture, whose congenital changes of intraepidermal or dermoepidermal adhesion induce the formation of blisters in the skin and mucous membranes, which may be spontaneous or caused by minimal trauma[1-2]. Its diagnosis is made from clinical and laboratory findings and, despite being a rare disease and with severe evolution, when the diagnosis is performed early, the family has a greater possibility of continuous monitoring, being referred to referral services that assist in care during the entire process of illness[1-2,4].

Although new cellular and molecular therapies are being investigated, there is no effective therapy or cure for EB and the treatment is symptomatic[5]. Thus, one of the main treatment measures is to prevent the development of new lesions, avoiding trauma, friction and pressure, especially in the areas of support and bony prominence[1-2,4]. Thus, the attention required by EB goes through skin care and lesions that include removal of crusts, dressings, drainage of bubbles and care in the bath. Thus, its complexity has an impact on the family, mainly because it is a disease with a high level of lethality in severe cases and that requires time and dedication to perform daily care[2,4,6-7].

Authors point out that the impacts of EB permeate the context of life of affected people and their families in different spheres[8]. They point out that the necessary performed care results in interpersonal, emotional, financial repercussions and sacrifices by parents and/or family caregivers. This implies the need for specialized centers and professionals prepared to support people with EB and their families.

Other authors corroborate and highlight that EB is a rare skin disease that requires support, care and continuous family attention[9]. In their study, they explored the experiences lived by family caregivers of EB patients and identified that family caregivers experience many challenges and experience stress and difficulties in caring for EB patients. The results indicate that parents experience a transformation and

impotence soon after the birth of a child with EB, and helplessness and lack of knowledge for being a rare disease, as well as the experience of adversities and challenges, which results in physical and mental exhaustion and also the need for support and social adaptation.

Although the stress of caring for a person with EB is experienced by the whole family, mothers are more involved in this process. Authors corroborate and highlight the mother as the main caregiver in the context of EB care[8-9], others show that the mother also has a prominent role in EB, since, most of the time, she assumes the role of the main caregiver, needing to reconcile this role with the other demands of daily life and own life[4,7,10].

In a study conducted by Wu and colleagues[9], more than 60% of family caregivers were represented by mothers. The authors highlight that they felt responsible for taking care of all rehabilitation, adaptation, growth and development of their children with EB, which can lead to overload with impact on the quality of life of these mothers, considering the need for constant care and the repercussions involved in the process of illness due to EB. In this regard, Dutch authors[11] point out that many studies address the quality of life of patients with EB, but that it is pertinent to study the relationship between parents and children and their coping strategies in the context of illness.

In this sense, when considering the mother as the main caregiver of the child with EB and recognizing the complexity of experiencing motherhood, from the birth of a child diagnosed with this disease, a rare condition that requires daily, continuous and complex care, the following research question arise: what are the coping strategies in the experience of motherhood regarding EB?

Thus, in order to contribute to the advancement in maternal and family care undertaken to children diagnosed with EB, as well as to subsidize health professionals, especially nursing, for family-centered care, based on the particularities imposed by this condition, this study aimed to know the coping strategies used by the mother of an infant with EB.

Method

This is a single case study research with a qualitative approach, structured based on the Consolidation Criteria for Qualitative Research Reports (COREQ)[12]. For being a rare disease with clinical dermatological and potentially severe evolution, the case investigated presented singularities that underlie the theoretical and methodological assumptions of investigating a phenomenon in its real context[13].

The case investigated in this study was identified through social media and selected for meeting the previously established inclusion criteria: to be 18 years old or older, to be a mother of a child under one year old with clinical or genetic diagnosis of EB, for being considered the most critical period for the performance of care, being a user of the Unified Health System (UHS) and residing in the state of action of the researchers, in order to enable data collection.

The collection of empirical data used as technique the in-depth interview and observation. The in-depth interview is understood as a conversation with intentionality, configuring itself as a flexible interview strategy, without the need for scripts and that seeks memories in a non-linear way, respecting the dynamics of the subject's narrative[14]. Thus, the guiding question "How has it been for you to experience the care of your daughter with EB?" was used to allow the gradual deepening of the participant's memories and narratives[14].

The fieldwork was carried out by meetings at the participant's home with previously scheduled dates and times, guided by her preference and by less interference in daily life, in respect of her privacy. Therefore, two meetings were held between February and March 2020. Soon after each of them, in order to complement and enrich the in-depth interview, observation records were made with the individual perception of thoughts, as well as intuitions of the researchers and manifestations of the participant, such as looks,

gestures, postures, silences, and the description of objects and scenarios[14].

For data organization, all the empirical material collected was entered in the Research Diary[15], which contains the transcripts of the in-depth interviews with the narratives in full and the observation records. The corpus of analysis of this study constituted a file, typed in Word, with 56 pages. Thematic content analysis was used, followed by the organization of analysis, coding, categorization and inference[16], which led to the emergence of two thematic categories: (re)meaning motherhood after the diagnosis of Epidermolysis Bullosa and the use of the internet as a resource for coping.

This study was evaluated and approved by the Research Ethics Committee of the *Universidade do Estado de Mato Grosso* (CEP/UNEMAT) under the registration CAAE: 26311819.2.0000.5166 and opinion number 3,779,764. The ethical commitment was mediated by the signature of the participant and the coordinating researcher of the Informed Consent Form, respecting the ethical precepts of research with human beings established in resolution 466/2012 of the Brazilian National Health Council[17].

Results

The studied case had as a center of observation and analysis the mother of a female infant with six months of age and clinical diagnosis of EB. The participant was a 26-year-old woman, brown, Protestant religion, university student, married and resident with the biological father of her only daughter, in a city in inland Mato Grosso-MT, Brazil. According to the mother's narratives, the infant was born from surgical delivery, with cutaneous aplasia in the right lower limb, in a hospital in the interior of the state, and was transferred to the reference hospital in the capital after seven days of hospitalization.

After the birth and diagnosis of the daughter, the mother reported that the support, especially from her parents and husband, was fundamental for the care of the infant. After discharge from the hospital, the mother recalled that she could only effectively assume the care required by her daughter one month after returning to her home. Based on the experiences of this mother, the analysis of the empirical material gave rise to the construction of two thematic categories, which present the coping strategies used by the mother in the experience of motherhood before the EB.

(Re)meaning motherhood after the diagnosis of Epidermolysis Bullosa

In the experience of the study participant, motherhood, before the birth of her daughter, had common characteristics and expectations of a pregnancy that evolved healthily and without complications. For the mother, the expectation was the birth of a healthy child, so that she could perform the care of the daughter immediately. However, this situation was reconfigured with the birth of the child with cutaneous aplasia in the lower limb and clinical diagnosis of a rare disease, as highlighted in her narrative:

> *At that moment I saw that motherhood had changed direction. Because we were waiting for a normal child, to put on her little earrings, put on her clothes, maternity clothes, which we bought months and months before. Totally changed.* [...]*We thought what will happen now? Will we be here tomorrow? Or not?* (Mother).

In view of what was exposed, the mother needed to (re)mean motherhood so that she could face the adversities imposed from birth by the illness of the daughter and those that would still be to come, in view of the lack of knowledge about EB. Thus, the mother recalled the 'change of course' in expectations regarding motherhood and how the clinical condition imposed specificities in motherhood after the birth of a rare child:

> *[Motherhood] changed direction. Because it was a way in which I had to* [be] *strong. I had to be even without it. I say it did not change direction negatively. It's just that we expected something, but we didn't have it. So, we moved on. We saw that there wasn't going to be a normal motherhood. But we tried to do it as normal as possible.* (Mother).

> *Then we saw how motherhood with a sick child is not easy. Thus, a normal child is hard work, with EB, it is three times as much work.* (Mother).

Regarding this period, the mother's memories about the birth and the first months of her daughter's life present themselves as difficult moments to remember. The photographic records, important to perpetuate memories in relation to motherhood, are directly associated with moments of uncertainty experienced by the mother, according to her narratives.

> *We don't see her photos. When she was little* [...] *we took a lot of photos. Like it or not, we were happy at that moment, because she was there. But when we see it, we feel sad. So, we don't see her photos from when she was a newborn*. (Mother).

Still in relation to these memories, the participating mother pointed out that she had the feeling of not remembering these difficult moments, experienced in the first months of motherhood, as a way of coping. Her narratives highlighted the barrier developed over time against the memories:

> *And I think that, I don't know if it's a defense, but it seems like I forgot that part [the first months of motherhood]. It seems I forgot it. It seems like I didn't live that part. Sometimes I think it's a defense, it seems like I jumped it. Sometimes I remember it when I see a photo, and all that bad stuff comes inside.* [...] *But when it's my normal day, it seems like I didn't go through it*. (Mother).

In the experience of this mother, the clinical diagnosis of a rare disease after the birth of her daughter imposed the need to (re)mean motherhood as a way of coping with the first adversities. After that, the mother had to deal with the lack of knowledge about the diagnosed disease in her daughter and seek alternatives to acquire theoretical knowledge about the condition, which is highlighted in the second thematic category.

The use of the internet as a resource for coping

The search for theoretical knowledge by the mother was the first way of coping with the situation experienced. This initiative aimed to support maternal and family care, which should be performed to the child. The narratives of the mother presented nuances of the way she sought information about the disease and the necessary care, in order to mean this moment, with the perspective of meeting the needs of the daughter. Thus, soon after birth and clinical diagnosis of EB, she realized that she would need to learn to care for the child from the needs imposed by the disease:

> *What I realized at that moment was what? She already had a diagnosis of EB. What was I supposed to do? Give her the best! I wouldn't regret it. Oh why does she have that?* [...] *I went looking for the best for her. There's no point in complaining, crying or acting like a poor thing.* [...] *Because I had to take care of her. I ran after everything, I ran after learning about medication, I ran after finding out about the best dressings they had, which reduced the pain* (Mother).

Faced with the need to learn how to take care of the daughter, from the access to theoretical knowledge, the mother sought strategies that allowed the contribution to this process. In this study, the participating mother used the internet as the main strategy to access information about EB. And from this, she built the theoretical knowledge that would subsidize child care, including later discharge to the family home:

> *After she was born, I studied a lot about diseases, medicines, what is best, what is not, creams, everything* [...]. *I read what EB was. I knew the theory a lot, right? I studied the disease a lot* [...]. *I researched and found out about the medicines* [...] *that gave me a better quality of life.* [...] *My mother came: [participant's name], what are we going to do? Which medicine to use? Because it was just us. In my city, no one knew about the disease, no one had knowledge.* [...] *So, we played the role of research[er], we ran after it. We saw that, to have her here today, we had to run after and fight*. (Mother).

In this study, from the constant searches on the Internet, over time, the mother was getting to know better about her daughter's disease and its repercussions. Soon, she realized the complexity of the disease and the high costs of the materials used for care, especially with daily dressings. Thus, in order to maintain her daughter's quality of life, she would also have to use the internet as a resource to build a social network of support and thus ensure the inputs used for the care of her daughter:

> [...] *Because, since she had this diagnosis, I realized that we had to give her the best. We always get the best treatments for her because we run after it, we fight*. [...] *We campaigned. She has a Facebook page, she has Instagram. We ran after international NGOs. Nowadays, we are part of international NGOs* (Mother).

> [...] *we started researching these dressings. Then the* [name of Brazilian NGO] *gave for us; after two and a half months, they arrived!* (Mother).

It is noteworthy that the mother did not find the necessary support for the care of her daughter with EB in the health system and network. Thus, the use of the internet and the search for Non-Governmental Organizations (NGOs) had established as an important coping strategy for care in all its dimensions. Including the search for knowledge, resources and the construction of a support network with people who have gone through the same situation and specialized institutions.

Discussion

In this research, the narratives of the mother highlighted the coping strategies used in the experience of motherhood after the birth of the daughter diagnosed with EB. It is known that the care required in this condition imposes specificities that need to be faced daily at home by the family and include wound care, hygiene, wound prevention, feeding, hydration, sleep, pain, heat control and itching[1,6,18]. Among the people who make up the family nucleus, the scientific literature highlights the mother as the main caregiver in cases of EB illness[2,4,10]. The mothers are mainly responsible for carrying out continuous care, being more exposed to physical, mental and emotional wear[2,8-9].

However, before the mother takes care at home, she needs to face the disease diagnosis process. In the course of motherhood, the idealization of uneventful childbirth and the birth of a healthy child is something that the mother expects, however, the arrival of a sick child can lead to the need to re-mean the conception of motherhood[19]. In the present study, the narratives of the participating mother indicated as the first coping strategy the need to (re)mean motherhood, so that she could face the adversities imposed by the rare disease diagnosed in her daughter.

It is noteworthy that this process causes suffering for the mother, because she needs to learn to re-mean all the expectations inherent in the process of motherhood[20]. Specifically in cases of EB, from the first hours of birth, the clinical evolution of the disease may impact, due to the extreme fragility of the newborn's skin[2,4,18]. In this sense, the mother participating in this study pointed out that the memories of the first months of her daughter's life present themselves as difficult moments to remember, considering that she was born with cutaneous aplasia in the right lower limb.

Another coping strategy that stood out as a finding in this study was the action of the mother to seek information about the rare disease on the internet. After the clinical diagnosis of EB, the participating mother realized that she would need to acquire theoretical knowledge about the disease in order to be able to perform the care required by her daughter. However, it is worth considering that, although the internet offers information that subsidizes families in the management of the care of children with chronic conditions, it has particularities that need to be analyzed during its use, especially regarding the reliability of the information accessed[21].

In this sense, the performance of health professionals stands out, especially those who make up the nursing team, in the guidance and preparation of the family for hospital discharge, as well as the indication of scientific sources for access and space for dialogue on the data found[18,21-22]. However, the literature shows that people and families who experience some kind of rare disease resort to health services and professionals seeking information and answers, and thus perceive the their unpreparedness to perform guidelines in relation to the condition experienced[23]. In this regard, health literacy is an important instrument of care and education in the area, since it corresponds to the ability of the person to access, understand and use information for the benefit of their health and well-being[24].

Particularly on EB, a recent integrative review highlighted the unpreparedness of health professionals regarding the guidelines on how the continuity of child care in the home should occur, and this led families to care in an empirical

and isolated way, without ties to health services and professionals[4].

Another finding worth mentioning refers to the use of the internet as a resource to build a social network of support, given the high cost of materials used to care for the daughter. A study conducted in inland Brazil highlighted the importance of the social support network built by the family to ensure the necessary inputs for the dressings and feeding of a young person with EB and also enable experiences of social interaction[10]. Corroborating this, a recent scope review carried out on the subject by Australian researchers highlighted that the concern with finances is one of the main impacts felt by the family experiencing EB[6]. This study pointed out that many parents need to reduce work hours or terminate the employment relationship due to the high burden of care required by their children and also deal with the expenses to buy the materials used in dressings.

In this sense, in order to guarantee these inputs by the UHS in Brazil, the National Commission for Technology Incorporation (CONITEC), in 2019, began the process to develop Clinical Protocols and Therapeutic Guidelines (PCDT) for EB, with contributions from society, associations and health professionals[25]. The PCDT was approved by the Ministry of Health (MH) on June 26, 2020 and defined criteria for diagnosis, treatment, monitoring and care lines in the UHS Health Care Network[26]. However, this document did not answer all the demands of individuals and families who experience EB, mainly because it did not ensure all the technologies recommended for care, leading societies and associations to contest the document[25].

Thus, after these mobilizations, the PCDT was updated in December 2021, with the inclusion of expanded information on non-drug treatment and the complementation of some sections of the protocol, such as specific and special dressings[1]. This technical document assures health professionals support to assist the person and family based on the current scientific evidence. It also highlights the possibility of access to the inputs necessary to perform care by the UHS.

Nevertheless, this study presents a limitation that needs to be considered, which refers to the difficulty of identifying people with EB, due to the absence of a national information system that aims to track them. Thus, it was possible to conduct a single case study and the narrated experiences refer to the experience of only one mother who has particularities to reside in the countryside. However, even with the limitations, this study contributed to the elucidation of coping strategies used by her to care for the infant daughter with a rare disease that requires complex care. By knowing these strategies, health professionals, especially nursing professionals, will be able to subsidize actions aimed at preparing the family to perform home care with technical and scientific support.

Final considerations

The study allowed knowing the coping strategies used in the experience of motherhood before the birth of the child diagnosed with EB. The present study highlighted that, although they were from health professionals, through the performance in the health care network, the competence to offer support to the needs of the family that experienced a rare disease, the mother needed to develop coping strategies independently. Her narratives presented as main coping strategies the (re)meaning of motherhood and the use of the internet to search for information about EB and resource to build and maintain the support network to subsidize the necessary care for the daughter.

In this sense, it is noteworthy that these results have the potential to enable health professionals, especially those who make up the nursing team, to recognize their role in the care of families who experience the child's illness by a rare disease. In care practice, the nursing team corresponds to the first support network accessed by the

mother and family and therefore needs to have knowledge and strategies to support them in coping with the particularities that involve the clinical evolution of the rare disease. Thus, to ensure comprehensive care in nursing care involves professionals' respect that the mother will go through a process of (re)meaning of motherhood and that the family should be prepared before discharge from hospital in order to acquire skills to perform child care at home.

Finally, it is evident the relevance of approaching the way the mother faces the process of illness and daily care in contexts of rare diseases. Especially about EB, currently, the focus of the scientific research is on the biological aspects of the disease and possible therapies. In this regard, it is essential to develop new studies in a similar context, since EB has different subtypes and the family experiences with the care given to the person with the disease can be different, and there may be other forms of coping, corroborating the scientific literature and the improvement of nursing care performed to the child and the family in the context of rarity.

Collaborations:

1 – conception and planning of the project: Ronaldo Antonio da Silva; Renata Emily da Silva dos Santos;

2 – analysis and interpretation of data: Ronaldo Antonio da Silva; Renata Emily da Silva dos Santos;

3 – writing and/or critical review: Ronaldo Antonio da Silva; Lidiane Cristina da Silva Alencastro; Renata Tomazelli Ferreira;

4 – approval of the final version: Ronaldo Antonio da Silva; Renata Emily da Silva dos Santos; Lidiane Cristina da Silva Alencastro; Renata Tomazelli Ferreira.

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