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Families' experience in managing children with sickle cell anemia: implications for care

Experiência da família no manejo da criança com anemia falciforme: implicações para o cuidado

Experiencia de la familia en el manejo del niño con anemia falciforme: implicaciones para la atención

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ABSTRACT
Objective: to understand families' experience in managing children with sickle cell anemia. **Method:** this qualitative study of 14 families of children with sickle cell anemia was conducted by semi-structured interviews addressing the families' management, experiences and coping from diagnosis to illness. The narrative technique was used and Symbolic Interactionism gave the theoretical framework**. Results:** the families' experiences revealed that they are constantly vigilant and protective, and have to face challenges from the moment their children are diagnosed with sickle cell anemia. Their progress was hindered by health personnel's lack of knowledge about the disease, lacunae in the health care system, and a lack of information about the disease, which imposed a reworking of family dynamics. **Conclusion:** health services and personnel must be trained to meet the needs and demands of children with sickle cell anemia and their families, and to provide information to strengthen them.
Descriptors: Chronic Disease; Anemia, Sickle Cell; Child; Family; Pediatric Nursing.

RESUMO
Objetivo: compreender a experiência da família no manejo da criança com anemia falciforme. **Método**: estudo qualitativo desenvolvido com 14 famílias de crianças com anemia falciforme, mediante entrevistas semiestruturadas abordando o manejo, as experiências e os enfrentamentos vivenciados do diagnóstico ao adoecimento. Utilizou-se como técnica a narrativa e como referencial teórico o Interacionismo Simbólico. **Resultados:** a experiência da família revela que ela permanece em constante vigilância e proteção, tendo de enfrentar desafios a partir do diagnóstico da anemia falciforme na criança. O desconhecimento dos profissionais sobre a doença, as lacunas na rede de atenção à saúde e a falta de informação sobre a doença marcam o itinerário, impondo a redefinição na dinâmica familiar. **Conclusão:** serviços de saúde e profissionais devem ser capacitados para atender as necessidades e demandas da criança com anemia falciforme e de sua família, bem como fornecer informações que as fortaleça.
Descritores: Doença Crônica; Anemia Falciforme; Criança; Família; Enfermagem Pediátrica.

RESUMEN
Objetivo: comprender la experiencia de las familias en el manejo de niños con anemia de células falciformes. **Método:** este estudio cualitativo de 14 familias de niños con anemia falciforme se realizó mediante entrevistas semiestructuradas que abordaron el manejo, las experiencias y el afrontamiento de las familias desde el diagnóstico hasta la enfermedad. Se utilizó la técnica narrativa y el Interaccionismo Simbólico dio el marco teórico. **Resultados:** las experiencias de las familias revelaron que están constantemente vigilantes y protectoras, y deben enfrentar desafíos desde el momento en que a sus hijos se les diagnostica anemia falciforme. Su avance se vio obstaculizado por el desconocimiento del personal de salud sobre la enfermedad, las lagunas en el sistema de salud y la falta de información sobre la enfermedad, lo que impuso una reelaboración de la dinámica familiar. **Conclusión:** los servicios y el personal de salud deben estar capacitados para atender las necesidades y demandas de los niños con anemia falciforme y sus familias, y brindar información para fortalecerlos.
Descriptores: Enfermedad Crónica; Anemia de Células falciformes; Niño; Familia; Enfermería Pediátrica.

INTRODUCTION

Sickle cell anemia is a common genetic disease in Brazil which leads to clinical manifestations such as pain crises, infections, dehydration, acute chest syndrome, severe anemia, neurological events and priapism[1]. The disease requires care maintenance which lasts for a lifetime, alternating between exacerbation and stability periods[2].

This disease is considered a chronic health condition and sometimes requires adaptations in the child's routine and their family as a result of complications. This is because they experience feelings of fear and insecurity in facing an unknown process which triggers expectations for information about the child's health condition and how the child's future will occur[1,3].

In this perspective, it is necessary for health professionals to not only be sensitive to clinical needs, but also to emotional, family and social needs, as well as to establish an educational process to assist the family

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in daily activities, health maintenance, prevention and control crisis, especially as this is a long-term condition[3].

Therefore, professionals working in primary care occupy a privileged place to offer support, in particular due to a previous bond established with the family and work developed in the territory[4]. However, studies point to gaps in this care process which directly reflect on managing children with chronic diseases, such as a lack of information about the disease, low resolution, and difficult access, among others[2,5].

It is believed that knowing the families' challenges as well as the way they manage the situation and deal with the child from the diagnosis time of sickle cell anemia can contribute to providing timely care and proposing appropriate interventions. Therefore, the guiding question of this study is: how is the management of children with sickle cell anemia performed by the family?

To answer the guiding question, the present study aims to understand the family's experience in managing children with sickle cell anemia.

THEORETICAL REFERENCE

Symbolic Interactionism (SI) guided the process of understanding the family's experience and its relationship with the influence of the child's chronic health condition, considering that this framework is a useful perspective for understanding human action since it focuses on individual human and group behavior[6].

This reference was chosen because the family is in constant symbolic interaction with its members, with the health team, with society, with the situations which emerge in their daily lives and in the interactional processes modified by the child's disease condition. The family attributes meanings in the social interaction with the experience which result from the interactions they have with the elements present in their experience with the child's disease.

METHOD

This is a descriptive study with a qualitative approach conducted in the hematology outpatient clinic of a large public hospital in a state in the Midwest region of the country. This service receives a daily average of 20 children from the capital and cities in the interior. Approximately 45 children with sickle cell anemia were being followed up at the time of the study.

Narrative was used as a research technique, which aims to extract, analyze and understand experiences, requiring the researcher to adopt analytical thinking to understand the narratives from a holistic and integrative perspective[7].

The guidelines of the Consolidated Criteria for Reporting Qualitative Research (COREQ)[8] guided the methodology description of this study. Families of children from one to twelve years old with a diagnosis of sickle cell anemia by hemoglobin electrophoresis was adopted as inclusion criteria. The initial age was established since the first symptoms of the disease usually occur at four months of life. Therefore, the family has already experienced several situations due to the disease by the time the child reaches one year of age. In turn, families who refused to participate (two), it was not possible to contact (three), or did not attend the service in the data collection period (26) were excluded.

The main researcher initially attended the outpatient clinic to observe the care provided by the team to the child/adolescent in order to get closer to the service dynamics in providing care to the children and families. For this stage, there was no previous script, however, the perceptions regarding the flow of care and the families' behavior were recorded in a field diary, which helped in the immersion of the data at the time of the analysis.

The main researcher presented the study and invited the families to participate approximately two months after the observation with the help of one of the service professionals. After acceptance, the interviews were scheduled to be held at the clinic before or after consultations in a private room or at the family's home, according to their preference. The participating families were selected by convenience from the days that the main researcher was present at the clinic.

Data collection took place from March to July 2016 through audio-recorded interviews which were conducted by the main researcher in an average of 2 meetings with each family, with an average duration of 40 minutes.

Each interview was conducted with care to allow families to retell their stories of child management and contexts imposed by sickle cell anemia. An initial script was used based on the request for a broad narrative of the family's experience in the context of the disease. New questions were asked as the narrative flowed in order to describe the

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experience. Examples of questions which guided the interview are: How has your family's trajectory been in managing healthcare for your child since the sickle cell anemia diagnosis? What has helped and what has made it difficult to treat sickle cell anemia? How have you been attending the services they use?

Data analysis followed the narrative research method with a category approach emphasizing content[7]. After the transcriptions and readings of the interviews, the significant texts which formed the categories were highlighted.

The study obeyed the ethical precepts and was approved by the Ethics Committee on Research with Human Beings (CEP) under opinion number 1,350,429. All participants signed the Free and Informed Consent Form in two copies, with the identification adopting the term "Family", followed by the interview number.

RESULTS AND DISCUSSION

A total of 14 families were interviewed, comprising 12 mothers, three fathers, two grandmothers, an aunt and a stepmother. Of these, ten lived in the state capital. The children's ages ranged from two to 12 years. Regarding the education level of the family member interviewed, three had higher education, one being incomplete, six with complete high school and five with incomplete elementary education. All children and adolescents were diagnosed at the time of neonatal screening.

The experience of families in managing children with sickle cell anemia is described from the representative theme of *The family in constant surveillance and protection*. In view of the diagnosis, the family is faced with a lack of knowledge about the disease and the impact that this event has on their daily lives, since it requires constant care for the child. Faced with a lack of information and uncertainties related to the disease, the family becomes vigilant and protective, starting to monitor the child and seek what they consider to be the best for the child's growth and development, as well as reorganizing themselves for the necessary confrontations. The categories which support the theme are described below.

From diagnosis to daily challenges

A sickle cell anemia diagnosis is impacting on the family, as it is not a known pathology which is disseminated in the population, even though it is the most common genetic disease in Brazil. The family's knowledge of the existence of the disease generally occurs with the first crises, the child's health worsens and there is a need for hospitalization.

> *Because we didn't want to believe that he had a disease. It took a while for us to understand. Because we saw that healthy little baby. Beautiful. He didn't present anything [...] We said "this is a lie, there's nothing". Because he didn't have anything. There was no crisis in the beginning. It was very difficult for us to believe. The day I really believed it was the day he had the first crisis. He was really ill [...] He has already been hospitalized. That was the day I really believed that this disease existed (Family 2).*

The initial ignorance of the disease prevents the family from seeking resources to reorganize themselves. When faced with the first symptoms of the disease, which are usually accompanied by the child's first hospitalization, the family starts to realize that something serious is happening, which leads them to believe that the diagnosis of the disease may be true. When searching for information, especially on the internet, the family is faced with images and texts which generate uncertainties, intensifying the anguish due to the possibility of the disease worsening.

> *At first it was very difficult. We didn't understand the disease very well [...] The first thing I did was read it on Google. Research about it. Then I was shocked, right? Because there it shows everything that is so bad that you can imagine (Family 4).*

Faced with a lack of knowledge about sickle cell anemia and the incomprehensible information they receive, the family seeks ways to understand the disease from data sources, often unreliable, generating greater apprehension.

Receiving information congruent with the demands of the family contributes to their understanding and enables identifying situations which may bring risk to the child. It is essential that health professionals propose health education actions that guide the family about this disease and its repercussions on the child's body. The correct information tends to avoid complications and favors the family understanding the illness process[3].

It is difficult for the family to deal with the uncertainties arising from the diagnosis, as they are unaware of their genetic inheritance and only become aware of the child's health condition after carrying out the neonatal screening test or as a result of the first crises in childhood.

This fact is a potential stressor for the family, since this chronic health condition will require constant adaptations in the roles and routines of family members in order to meet the care needs. These findings corroborate the authors of

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a study which recognizes the repercussions of the disease on the child's and family's lives and on the interference they cause in family dynamics[9].

The mother usually takes care of the sick child. In the context of sickle cell anemia as a chronic disease, this situation can cause conflicts in the marital relationship, with breaking ties and care overload for one of its members[10]. The responsibility of a family member for the event, added to the lack of knowledge about it, favors breaking family bonds:

> *For me it's difficult, because when I found out that D. had sickle cell anemia, his father didn't want to be with us anymore. He said he was leaving, because I was guilty, everything came over me (Family 9).*

The chronic disease situation reinforces the role of caregivers assigned to women - mothers and grandparents. The care of children with sickle cell anemia is often centered on the maternal figure, who in many cases does not receive support, generating overload and emerging anxiety, depression and social interaction problems[11]. Thus, it is noted that the crises of the disease disturb the family order and routine, as well as being factors of uncertainty which affect their lives and can potentiate other conflicts which already exist in the family.

Family coping

The coping process begins with the discovery of the disease and its impact on the family, which seeks to assign meanings and mobilize resources such as religion and faith for the necessary support in the family's daily life with the child. The child's suffering caused by pain crises leads the family to strengthen their bond and their spiritual belief:

> *Certainty is faith, it is believing in God, which is a support that gives us the structure to go through it. Because there are days, really, when you have a person with a chronic illness in the family, you don't want to get out of bed, you don't want to see the person you love suffering so much (Family 13).*

It is essential that the health team knows the religious beliefs of the families, thereby enabling their spiritual care which helps in coping and strengthening resilience. There is evidence that religiosity and spirituality are inherent components of coping with diseases, being important resources for family members, especially in the face of a threatening prognosis[12].

The family emphasizes that love is a great motivator in searching for better conditions for the child, in addition to strengthening family resilience. The child's inner strength strengthens the family in facing their challenges, while the fear of losing the child triggers a mobilization of resources and to struggle for them.

> *It may sound silly, but it is love. We have such a great love for him. He is such a special kid, that it makes us fight more. The fear of losing him is so great that we fight more. We fight, we do what we can (Family 14).*

Knowing and living with the child's disease contributes to better coping by the family. Being with other parents who have similar experiences helps families to exchange experiences and share information which helps them in their daily lives and in protecting the child.

> *Knowing other cases helped a lot. Other than that, […] it's day-to-day. Day after day, we learn to live together. The coexistence I had with other parents during the hospital stay helped me (Family 4).*

The support network set up by other parents living in similar contexts enables the families to share experiences and develop bonds which strengthen and help them. Family participation in social groups of people with sickle cell anemia provides an exchange of knowledge and common anxieties, promotes strengthening and improves coping with the situation[13].

The family feels the need to be guided about what is happening to the child so that they can act and feel safe in handling the situation. Having access to information minimizes family anxiety:

> *What helped was to have more explained, we have more information so we don't get anxious, because the internet said that (Family 3).*

It is necessary to know how to cope with, care and protect, to know what the signs and symptoms are, what can be done to prevent the child's crises, as well as to know how other families handle similar situations. In this sense, the family becomes responsible for the treatment routine, self-care and for the perception that the child and the family themselves have about the chronic condition[11].

The family takes protective measures and care in order to protect the child and prevent new crises of the disease, which in turn prevents the child from having a common life and playing with friends. Fearing aggravation of the disease, it keeps the child away from common events in childhood.

> *They always asked me a lot to bathe in the rain, I never let them, I know they were so suffocated, they felt suffocated. Because, I didn't let them eat industrialized food, just healthy things. I didn't take them to a*

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> *birthday party to avoid eating cake, sweets, as I thought if I overprotected them, they wouldn't have crises (Family 13).*

The fear of crises or worsening of the disease produces over-protection and reframing of childhood by the family of something normal for this stage of life, making it as something fearful and uncertain. Overprotection refers to how family members perceive the child's health weaknesses. The child's symptoms can potentiate the family's concern[14] so that they can be over-protective due to the family's difficulty in solving situations which cause distress.

In the itinerary in searching for information and care for the child, families also face professionals' lack of information and the invisibility of the disease, manifested by their lack of preparation to recognize the characteristic signs and symptoms of sickle cell anemia, treatment and crisis management.

Faced with the professional approach and unpreparedness in caring for the child in crisis caused by the disease, in addition to reassuring the child, the family needs to convince the professional about the severity of the child's illness and instruct the conduct that they must take:

> *My daughter screamed in pain when I arrived at the hospital, I told the doctor that she had sickle cell anemia and that dipyrone was not going to solve it, but she ignored it and my daughter was screaming for over an hour (Family 13).*

The reference services are configured as an important support for the family, because they are where they get some of the necessary information for providing care for the child through professionals trained to care for the child. However, they do not find help in emergency situations for the child, especially in specialized services, since they work on an outpatient basis and through scheduling appointments.

A study which discussed the (in)visibility of children with special health needs and their families revealed a kind of transfer of responsibility for the care and monitoring of these children to specialized institutions, which has repercussions on the lack of care demands of this group in other healthcare services. Thus, a guarantee of care for these children is a challenge to be overcome by families, who have not ensured adequate continuity of the child's health follow-up[15].

Of the 14 families interviewed, nine do not access services other than the reference hospital and the specialized outpatient clinic, as their experience with other services shows that professionals are unaware of the disease and the appropriate therapeutic conduct, and therefore they perceive an unpreparedness for managing crises in children with sickle cell anemia:

> *I told the doctor that she had sickle cell anemia and the doctor asked me how she got this disease (Family 13).*

Health professionals' lack of knowledge about the disease perceived by families during the care of the child transmits a feeling of invisibility of the disease and of not validating what they are experiencing. It is worth noting that the healthcare situation provided to people with sickle cell disease is precarious, since health professionals are unaware of the peculiarities of monitoring, leading families to not recognize primary care as a place to take care of various aspects of their health[2].

Families feel the need to be monitored and better informed for decision-making, including about future family planning.

> *When the doctor spoke, he spoke, but he did not give incisive attention to explaining how it is [...] He just said that he couldn't do it anymore. But, he did not explain what the disease was like. Because I think that if I had it explained to me in depth, we wouldn't forget it so easily, right? So he told us to avoid [...] it, right? Having a child. But time passed and eight years later we forgot (Family 8).*

The family narrative reinforces the importance of receiving clear and detailed information, especially regarding risks. In this sense, the Ministry of Health points out that the guidelines must be transmitted in appropriate language and with sensitivity, as the emotional moment of the family may not favor acceptance and immediate understanding of the content. In addition, it recommends genetic counseling, addressing hereditary aspects and other contexts of the disease as an essential component of the conduct of health professionals who provide care to children with sickle cell anemia and their families[16].

Study limitations

As a limitation of the study, the impossibility of generalizing the results due to the performance in a service with specific characteristics stands out.

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CONCLUSION

After diagnosis, the families of children with sickle cell anemia have difficulties in understanding the disease severity and the importance of clinical stability in the first months of life. They find strength in faith, in spiritual beliefs and in the child themselves to continue in the search for better conditions for their health and growth in order to cope with the disease.

Faced with the constant challenges imposed by the disease, the family handles and copes with the situation as best as they can, but they do not always find the necessary help or support from health professionals. Professionals in specialized services represent an important source of information for the family, but access to them is punctual and during consultations and exams, which makes it impossible to clarify doubts about sickle cell anemia and the child's health. Therefore, it is essential that health services and professionals are trained to meet the needs and demands of children with sickle cell anemia and their families.

The importance of clear and appropriate information is highlighted in this study in order to qualify the care of children with sickle cell anemia and their families. Attentive listening, offering adequate treatment and providing guidance on the disease and its implications for the child's and family's lives are relevant to produce a paradigm shift in the care of this population.

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