

The experience of disease in cystic fibrosis: the paths to comprehensive care*

A EXPERIÊNCIA DA DOENÇA NA FIBROSE CÍSTICA:
CAMINHOS PARA O CUIDADO INTEGRAL

LA EXPERIENCIA DE LA ENFERMEDAD EN LA FIBROSIS QUÍSTICA:
CAMINOS PARA EL CUIDADO INTEGRAL

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ABSTRACT

Cystic Fibrosis is a chronic disease with an extensive impact on family life. The experience of disease is the manner how individuals respond to the disease, assigning meaning and searching for ways to deal with it in their daily lives. The objective of this study was to understand the experience of Cystic Fibrosis in the family context. This is an ethnographic study, performed with families of children assisted at a teaching hospital located in the state of São Paulo. Results were divided into the themes past, present and future, and all phases were permeated with the search for the meaning of the disease and social support, the importance of religion and spirituality, and child's socialization. Knowing the experience in the disease and the social network is indispensable when planning comprehensive care. This is an innovative approach in health care for chronic diseases.

DESCRIPTORS

Cystic fibrosis
Child
Family
Pediatric nursing
Religion

RESUMO

A Fibrose Cística é uma doença crônica que grande impacto exerce sobre a vida familiar. A experiência da doença é a maneira pela qual os indivíduos respondem à doença, atribuindo significados e buscando maneiras para lidar com ela no seu cotidiano. O estudo teve como objetivo compreender a experiência da Fibrose Cística a partir do contexto familiar. Estudo de caso etnográfico, realizado com famílias de crianças com a doença atendidas em um hospital escola do interior do estado de São Paulo. Os resultados foram divididos nos temas: passado, presente e futuro e permearam essas fases a busca pelo significado da doença e pelo suporte social, a importância da religião e espiritualidade e a centralidade da socialização da criança. Conhecer a experiência na doença e a rede social torna-se imprescindível para o planejamento de um cuidado integral. Essa abordagem mostrou-se inovadora para o cuidado à doença crônica.

DESCRITORES

Fibrose cística
Criança
Família
Enfermagem pediátrica
Religião

RESUMEN

La Fibrosis Quística es una enfermedad crónica que ejerce gran impacto en la vida familiar. La experiencia de la enfermedad es el modo en el que los individuos responden a la enfermedad, atribuyendo significados y buscando maneras para lidiar con ella en su cotidiano. El estudio objetivó comprender la experiencia de Fibrosis Quística a partir del contexto familiar. Estudio de caso etnográfico, realizado con familias de niños con la enfermedad atendidos en hospital escuela del interior de San Pablo. Los resultados se dividieron en los temas: pasado, presente y futuro y atravesaron esas fases la búsqueda del significado de la dolencia y el apoyo social, la importancia de la religión y espiritualidad y la centralidad de socialización del niño. Conocer la experiencia de la enfermedad y la red social es imprescindible para el planeamiento de un cuidado integral. Este abordaje se mostró innovador para el cuidado de la enfermedad crónica.

DESCRIPTORES

Fibrosis quística
Niño
Familia
Enfermería pediátrica
Religión

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INTRODUCTION

Cystic fibrosis (CF) is the most common genetic chronic illness in the white race⁽¹⁾ affecting one in every 2,500 liveborns in Europe, with similar incidence levels in Brazil⁽²⁾. This congenital and multisystemic disease affects the exocrine glands and can occur in different epithelial cells, including sweat and pancreatic ducts, airways and biliary ways, intestine and deferent duct⁽¹⁾.

CF treatment aims to keep the lungs clean through aerosols and respiratory physiotherapy, and also to maintain good nutrition, including nutrient and pancreatic enzyme supplements. In case of infection, antibiotics are needed, often demanding patients' hospitalization⁽¹⁾.

The discovery of the CF gene in 1989 permitted the establishment of treatment coherent with physiopathology, contributing to increased life expectancy, which in developed countries figures around 40 years⁽¹⁾. A significant improvement in life expectancy occurred in comparison with the 1940's, when 70% of cases died before their first birthday⁽³⁾. In Brazil, life expectancy ranges around 18 years, a fact that has been justified by the delayed diagnosis⁽⁴⁾.

Because this is a systemic disease that is complex to treat⁽¹⁾, the family needs to be involved in care, especially in case of children and adolescents. As treatment occurs daily and several times per day, a caregiver delivers care⁽⁵⁾, frequently the mother⁽⁶⁾.

Most health actions focus on the individual, while the family is seen as a complement⁽⁷⁾. Understanding the way the family, including the individual, experience the disease gains relevance though, in order to understand the strategies used to cope with it and understand it.

The disease experience is the way people place themselves and assume the position of being ill, giving meaning and finding routine ways to deal with the situation. The way they respond to the problems deriving from the situation are socioculturally constructed and remit to a world of shared beliefs, values and customs, just like the meaning itself of the disease. As experience is gained in the social sphere, it is also intersubjective and it is in daily life that people construct actions together to deal with the disease⁽⁸⁾.

The disease is a problem situation that affects life and requires restructuring from the stakeholders. As this is a situation that generally reveals a lack of knowledge, it mobilizes people to seek new practices in order to deal with and explain the problem⁽⁹⁾.

Although the biomedical model provides information about the disease, through objective diagnostic methods, this explanation alone, when assessed from the patient's viewpoint, does not manage to answer the questions in-

involved in the disease process, such as: Why me? Why now? These questions seek the cause of the disease and its meaning. The construction of these answers often demand meanings and beliefs from the sociocultural context⁽⁹⁾.

Chronic childhood illnesses demand that the health team adopts different management and postures towards children and their families. Questions inherent in care quality and this population's life projects should be granted more relevance, as well as the cognitive and emotional aspects that influence decision making to cope with the health problem, as well as its evolution and treatment adherence⁽¹⁰⁾.

The approach of the disease experience permits acknowledging important dimensions of the disease and treatment, so as to reach a broader understanding of the disease phenomenon. Through the narrative, the disease experience is expressed, interpreted and communicated, normally in a time period of actions, and contains knowledge gains from past experiences, projecting them into future actions. In the narratives, suffering takes form, allowing people to organize their experience and transmit it to others⁽⁸⁾.

Thus, the aim of this study was to understand the disease experience of Cystic Fibrosis based on the family context.

METHOD

This paper was taken from the Master's thesis entitled *Living with Cystic Fibrosis: the disease experience in the family context*. This qualitative ethnographic case study⁽¹¹⁾ involved families of children with Cystic Fibrosis attended at a teaching hospital in the interior of São Paulo State.

The ethnographic case study was used because it permits the comprehensive analysis of phenomena, from the subjects' viewpoint, starting from the interaction between the researcher and the research subjects⁽¹¹⁾. Ethnography proposes studying people in their natural environments, with a view to apprehending social meanings and daily activities, which demands the researcher's participation in the environment, turning him/her into the instrument of the ethnography. Hence, culture represents the fundamental theoretical premises, based on which data will be extracted and interpretations will be developed⁽¹²⁾.

In health, ethnography permits describing health, disease and care, considering the broad and complex universe of social relations, that is, in which the understanding of these concepts and health practices constitute the general picture and that of the social order⁽¹²⁾.

Data were collected between April and November 2007. Participants were captured at the University of São Paulo at Ribeirão Preto Medical School Hospital das Clínicas - HCFMRP/USP. This is one of the seven treatment centers for the disease in São Paulo State. Today, it attends 84 pa-

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tients, including adults, adolescents and children, most of whom (28 %) of preschool age (between 3 and 6 years).

To get to know the disease experience in the family context, the place chosen for the study was the environment where the phenomenon takes place⁽¹²⁾. Fieldwork occurred in those place the family indicated as the locus of the events. The home was considered the starting point and most representative place of family life. Departing from the home, the family indicated other places of reference: school, work and neighborhood.

Approval for the study was obtained from the Institutional Review Board at the University of São Paulo at Ribeirão Preto Medical School Hospital das Clínicas in February 2007. To guarantee anonymity, respecting the orientations expressed in the Informed Consent Term and the participants' desire, families were identified by order of data collection (family 1, 2 and 3) and received fictitious names.

Inclusion criteria were: families who had children with Cystic Fibrosis under follow-up at the institution for at least one years and being considered a critical case by the multidisciplinary Cystic Fibrosis team.

Hence, sampling was intentional. Among intentional sample types, the critical case sample was chosen, with cases that are considered dramatic or, for some reason, particularly important in the study context⁽¹¹⁾. Critical cases were considered as cases that presented important issues for the family dynamics and disease management, such as: adopted or abandoned children, families going through the separation process, disease considered severe.

Study participants were three families. The first family was chosen because it was nuclear, in which CF became the center of activities, creating marriage conflicts. In the second family, the child is the main caregiver's great-granddaughter and was rejected by the biological mother due to the disease. In the third case, the family adopted the child with CF.

Among the relatives, two mothers participated, one great-grandmother, one great-grandfather, one great-aunt, one father and two sisters, with ages ranging from 18 to 80 years, all of them living in the city where the children receive treatment. The children were two girls and one boy, all of them five years old and attending school regularly. The two girls, from families 1 and 3, have a severe form of the disease, according to the multidisciplinary team's criteria, while the boy from family 2 has the moderate form

Data were collected through seried open interviews and participant observation, which synchronically composed the field work⁽¹²⁾. Upon the first visit, the genogram and family ecomap were constructed and the guiding question was presented: *Tell me about the history of CF in your life and in your family's life*. On average, each family received five visits.

Data analysis started with the transcription of the empiric material, in the attempt to build a text with the inter-

view data, in combination with data from participant observation. To systemize the process, the scheme was used for qualitative data analysis, including the process of notifying, joining and thinking. This is a spiral-form process, that is, the phases are not linear and ordered. The author classifies it as interactive and progressive, as it turns into a cycle and repeats itself. It is also recursive, as one phase can make the researcher return to the previous one, and holographic, as its part of the process contains the process as a whole⁽¹³⁾.

So as not to break up the text and use the analysis strategy without impairing the contents as a whole, the data mapping strategy was used, schematically joining the data⁽¹³⁾. The transcribed material was printed, with a wide margin on the right where guiding notes were made, after reading the material line by line. After data systemization, they could be coded, i.e. seeking phrases, discourse, words, impressions that were similar or had the same meaning across the material. Colors were attributed to each code for coding purposes, based on which themes emerged.

RESULTS AND DISCUSSION

As described earlier, the disease experience is something fluid, continuous, practical and processual⁽⁸⁾. Thus, the researchers chose to present it based on the timeline: past, present and future.

Past

The study participants started the narrative by remembering the start of the disease. The past presents this phase on the timeline. The start was related with a crisis period for the family, due to the difficulty to reach a diagnosis and the severity of symptoms:

One day I arrived to visit him and the doctors came to explain the disease, because I didn't know what it was, they said it was Cystic Fibrosis, that there was no cure, that it was very severe, it seemed as if the whole hospital had fallen on my head. I got so desperate that I kept on talking, a lot of physicians came to talk to me, the psychologist and over time I calmed down... (Nadir, great-grandmother - Family 2).

The delay to diagnose CF can contribute to worsen parents' negative feelings and guilt, and also cause less confidence and hostility towards the health team⁽¹⁴⁾. We identified a feeling of guilt and fear of generating a new family member with the disease; these data were also found in other studies involving parents of children with CF^(5,7,14).

At first I thought: I had so many opportunities, so many people I dated, almost married... why on earth did I choose my husband, he who has this too (Camila, mother - Family 1).

When a stressful event like the disease becomes part of people's life, it needs to be integrated into the scheme of daily life. As a part of this integrative process, further knowledge on the problem is needed and, hence, meanings need to be attributed, including that of cause⁽⁸⁾.

Explanations on the causes of diseases result from the inter-relation of elements in the sociocultural context, which can be included in three large categories: causes related to natural factors, to supernatural factors and to personal factors. For causes related to natural phenomena, a causal agent is not identified. Supernatural causes include religious and spiritual explanations, while personal cause relate the disease with phenomena from the patient's social life⁽⁹⁾.

At first I asked myself: My God, what is it me and my husband did that was so severe that my daughter has to pay for it? I got revolted with God and didn't event pray anymore (Camila, mother - Family 1).

He was born with this, it was God's will... He wanted it that way and it had to be him [Guilherme]... If not, why didn't his brother get it? (Véio, great-grandfather – Family 2).

Supernatural and personal explanations were also observed more frequently than natural ones in other studies⁽⁸⁾. That is the case because, in view of the diagnosis, besides the biomedical explanation, meanings are required that relate the disease with the order of the world and with the social order the person is inserted in and this search takes place in other cultural spheres, like in religion⁽⁹⁾.

The families expressed the lack of orientations about the disease, provided by health professionals, especially at the time of the diagnosis. They also mentioned other forms of getting these orientations, although they were not always sufficient to reach an understanding:

Until today I cannot explain exactly what it is, it's really difficult, but at first we had no idea... first because his mother didn't tell us, second because the doctors didn't call us anymore to explain it like, really WELL [emphasizes the word]... (Márcia, aunt- Family 2).

I went to the Internet to see what the disease was. Dear God, I got even more desperate! It just showed bad things... I even got rid of the Internet at home so that I wouldn't see it anymore (Camila, mother-family 1).

The way the family is supported at the time of the diagnosis and the explanations received exert important influence on the way they will cope with the disease. The information received during this period is marking and taken along over time. Thus, the diagnosis period plays a determinant role, as it will be reminded across the disease experience⁽¹⁵⁻¹⁶⁾.

The interpretation people elaborate for a given experience with the disease, such as its emergence, results from the various means through which they gain medical knowledge. This knowledge differs among people and is permeated with subjectivity. The interpretation is also continuous and can change, as the disease experience is temporal and alters like the disease, and also because this knowledge is constantly confronted in daily life, either by family relations, relations with neighbors, health professionals and friends. Thus, more than one explanation for the disease will be constructed, as knowledge is recurrent, process-based and contextual^(8-9,17).

After the diagnosis, family restructuring required the search for instrumental support. The family's help and the search for support from other family members were mentioned, although the extended family helped little in care, posing an overload on the main caregiver:

It's very difficult, I won't lie by saying that it's easy, because it's not. I wake up at 04:30h in the morning. I make my son's lunch, the boys' father, because he goes to work at 6h. Then I prepare Guilherme's things, give his medicine and make him ready for school, because he starts at 07h. Then I put the house in order, clean everything, because neither I nor Guilherme can handle dust and then I make lunch. So, at noon, I'm already dead tired! (Nadir, great-grandmother- family 2).

Other studies on the impact of Cystic Fibrosis, involving families, mentioned the family's overload, as she is normally the main caregiver, as a preponderant factor for stress due to care delivery to a child with Cystic Fibrosis, making her lose the functions she used to perform, such as work for example^(15,18).

Although the results are organized according to time phases, the disease experience and its course occur in a continuum. Therefore, changing from one phase to another does not mean that issues present in the previous phase will be forgotten, but accumulated, matured, reconsidered and reviewed⁽⁸⁾.

Present

After the diagnosis phase, the study participants attempted to review the meanings attributed to the disease, not only by family members but also by the children themselves. Through the new experiences, the disease is resignified, although the religious conception is still present to explain it. If, in the diagnosis phase, the disease was seen as a punishment, in the present, it is understood as something beneficial, as exemplified next:

The disease is a mission the family has to fulfill... it was God's choice, we are special, she's special. The disease came to teach us, you know, for us to gain maturity... (Camila, mother - Family 1).

Supernatural causal explanations appeared as a possibility to associate the disease with something positive and, thus, seek forces and hope to deal with the CF and with the obscure future, glimpsed in the diagnostic phase, in accordance with a previous study⁽¹⁵⁾.

Religion and spirituality continue as spheres that underlie the disease experience in family life and, now, with the children growing up, they start to be inserted in these realities. Although the reason to seek spiritual and/or religious support has changed, the search to find a meaning for the disease and to maintain the child's health continue as central themes:

Now he gets his medicine himself and takes it, I don't have to keep on repeating it. He got new milk at the hospital, but at first he didn't like it much. Now he gets it himself, shakes

and drinks it, the way the nutritionist explained him, then he says: I'm gonna drink it because I want to get strong. I'm gonna take this medicine please Jesus, cure me God! (Nadir, great-grandmother – Family 2).

The search for God's help in times of anguish remits to a supreme power, who can do anything. Religion, sought as a form of support, is a sociocultural conception, apprehended in the family sphere and in the social group the child is inserted in. In this sense, the doctrine followed is not important, but the type of support religion offers.

Interest in the importance of knowledge on the disease's dimensions for the child is increasing, including religion and spirituality, which have shown to be relevant and crucial for their wellbeing, especially in view of stressful situations like hospitalization, disease and death⁽¹⁹⁾. Religious systems permit an interpretation of the disease that inserts it in the broad sociocultural context of the person holding that interpretation. More than relating the disease with a specific cause, religion permits the organization of the states of confusion and suffering the patient experiences^(12,17).

The families in this study showed a trend to overprotect children with Cystic Fibrosis, as a form of compensation for the crises and losses the disease entails:

I make her do everything they say that will do her well, she does ballet, tap dancing, swimming in school... Also, when she's not that well we let her eat what she wants, because we feel sorry, we know that it's not right, but we compensate a bit for her suffering (Camila, mother- Family 1).

Although the parents sometimes compensate for the disease through overprotective attitudes, this is not recommended to other people as, according to the mothers, they do not want the children to feel different from others:

There's no difference because I said right at the start that she's a normal child, you have to fight, punish them like other children, the only thing different is the food, the enzymes... (Marinês, mother - Family 3).

Overprotection can emerge as one of the consequences of the feeling of guilt, of uncertainties on the child's development and fear of death. It should be highlighted though that there is a thin line between overprotection and answers to the sick child's true needs⁽²⁰⁾.

As the children grow and are included in the social context, new support networks are developed for socialization and health care. During this period, the choice of environment, such as school and friendship networks, is weighted based on their acceptance due to the CF:

I chose the school after talking to the director. I explained what the disease was and asked if there was any problem for her to study there. She was very nice and said that didn't have anything to do with it, so she studies there until today. Oh dear... they help me a lot there, they give the medicine correctly, it's great! I'm even afraid for when she'll have to change (Camila, mother - Family 1).

School-age children with chronic illnesses are at the intersection of health and education system, as they are separated at teaching institutions in most countries. Thus, these children have needs that are not fully attended to, creating deficiencies in one of the sectors, generally the educational, in view of the urgency of health⁽²¹⁾. In that sense, the lack of communication between these sectors is the main educational obstacle children with chronic illnesses face⁽²²⁾.

When considering chronic childhood illnesses, one relevant care aspect relates to maintenance or non-impairment of the child's growth and development due to the disease⁽¹⁰⁾. Hence, thinking about the chronic illness from the child's perspective becomes particularly valuable, as it permits understanding part of this childhood experience, from the perspective and in the environment characteristic of this phase of life. In those cases when social support, particularly school, did not show to be effective, it was perceived that the family, including the child, chose to hide the disease from other people:

If people knew, I think there would be a lot of prejudice, I don't tell, even the teacher told me not to tell, she said like 'he's special and I like him a lot, but if the other children find out they're gonna make fun of him' so we don't tell (Nadir, great-grandmother - Family 2).

It is at school that the disease is exposed and, sometimes, that the child with Cystic Fibrosis perceives that, in fact, (s)he is not like other children, mainly in terms of daily routines, due to the medication and disease symptoms⁽²²⁾. When the school environment cannot bear the differences, it can turn into a space where stigma is perpetuated and maintained⁽²¹⁻²²⁾. As ways to cope with stigma, besides covering up, patients can start to neglect treatment in the school environment and, consequently, besides stigma itself, the disease may worsen⁽²¹⁾.

On the other hand, when school is an effective part of the family's instrumental support network, the child's socialization, as well as treatment, is accomplished without difficulties in this sphere. When teachers, relatives of classmates and students themselves know the disease, not only do the child's socialization and education occur naturally, but health care is shared.

Future

In the study participants' narratives, the future was interpreted as the temporal environment of continuity, of the family and Cystic Fibrosis patient's permanence in the objective social world in a way similar to or better than what happens at present. Therefore, the participants related present with past attitudes:

We take good care of him because, in the future, who knows there might be a cure. We have to do things always thinking about the consequences further ahead, because there's no way to turn back later, just regret... (Márcia, aunt – Family 2).

In previous studies involving children and adolescents with CF and their relatives^(6,22), the hope placed in the fu-

ture showed to be necessary to keep up health care, and also to stimulate life, for patients as well as for people sharing the disease experience.

In the formulation of future projects, actions are assessed, considered and projected and the issues highlighted in the present study narratives, such as the network and social support, stigma, religion and the meaning of the disease, are launched into the future in order to seek positive experiences. When reassessing past and present events, the relatives expressed inquiries about improvements they could gain in the future, related to the strengthening of social support networks:

At first, when we discovered what she had [Cystic Fibrosis], we got desperate and didn't really know what it was... The support group that exists now did not exist. I think this group should get stronger, you know? It would help the mothers more. GAFFIC [Support Group for Family Members and Patients with Cystic Fibrosis] is very important, but it's very difficult, you know... (Camila, mother – Family 1).

CONCLUSION

In this study, the goal was to understand the experience of living with CF based on the context of families that include children with the disease. In this sense, the intent was to contribute to the way care delivery and health care for children with Cystic Fibrosis are taken into account, including the consideration that the family and the community share the disease experience.

Health care for children with chronic illness has been fragmented and, until today, based on exclusively biomedical aspects of the disease. Thus, acknowledging the disease experience as a practical approach is elucidating, as it permits care planning in a less segmented way, keeping in mind aspects of the disease and chronicity process that are not included in other approaches but are very influential in treatment management, in the signification of the disease and in the child and family's growth and development processes.

The family shares its experience with the disease to the extent that it attributes meanings to the events and seeks support in the social network. The need appeared for further analysis of communities and the way they compose

health care, together with family members and the medical care system. It is only based on knowledge about these support systems and their functioning that communities' inclusion in health care can be considered and put in practice.

Religion emerges as an element that permeates the people's and families' disease experience, turning it into an important coping resource. It comprises the sociocultural system that signified the suffering during the studied families' trajectory, and thus provided the emotional support that was so often demanded. Acknowledging the importance of the religious and spiritual sphere in care planning and therapy assessment showed to be fundamental for a respectful approach to these patients.

Likewise, recognizing the meanings attributed to the disease is essential for care planning, to the extent that it is by attributing meaning to the disease that the search for help, treatment and its assessment will be accomplished. As religion and spirituality mediated a large part of disease signification, getting to know and including these in daily practice gains importance. Spiritual and/or religious care is particularly relevant for nursing, as it is present at times when this support is needed, such as the diagnosis, crises and death. For this care to be delivered adequately, first, it needs to be acknowledged, as well as the patient's desire to receive it.

In view of the Brazilian reality and relating chronic illnesses and school, nurses can be of help with educative measures and interventions in schools where children with CF are present. Through health education and care planning at the school, issues related to disease stigma and treatment adherence can be detected and attended better.

The above reflections raise questions regarding comprehensive care, including primary health care for chronic conditions. Hence, it should be considered what innovative care is being proposed to address these conditions and, as specialists, how comprehensiveness can be added to daily practice.

As an ethnographic case study, generalizations are not possible but, in view of the sociocultural nature of the study phenomenon, reflections are possible on the central position of the disease experience in the health-disease process and on care delivery in chronic conditions.

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