



ISSN: 2230-9926

Available online at <http://www.journalijdr.com>

# IJDR

International Journal of Development Research

Vol. 11, Issue, 11, pp. 52183-52185, November, 2021

<https://doi.org/10.37118/ijdr.23409.11.2021>



RESEARCH ARTICLE

OPEN ACCESS

## EXPERIENCES OF FAMILIES WITH CHILDREN WITH CYSTIC FIBROSIS IN THE FAR NORTH OF BRAZIL

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### ARTICLE INFO

#### Article History:

Received 14<sup>th</sup> August, 2021  
Received in revised form  
07<sup>th</sup> September, 2021  
Accepted 05<sup>th</sup> October, 2021  
Published online 30<sup>th</sup> November, 2021

#### Key Words:

Children, Cystic Fibrosis,  
Family Experience.

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### ABSTRACT

Cystic fibrosis is a rare, chronic, progressive and disabling disease, imparting hardship not only to the patients but their families as well. Family resilience within the context of cystic fibrosis is evidenced as an intrinsic and interrelated process, requiring a sensitive and broader look at the object of study. This study examined how families are restructured around the challenges presented by cystic fibrosis. A descriptive qualitative approach guided by the methodological theoretical framework of Symbolic Interactionism and Narrative Research was used. The analysis of the narratives resulted in the identification of two themes: The family and the child living with a new routine and Resilience in the trajectory of families. It concluded that the crises and difficulties caused by cystic fibrosis promoted the development of skills that favored family resilience.

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Citation: Ketlen de Sena Silva, Caren Julianne F. de A. Mello, Anneli Maricielo Cárdenas Celis et al. "Experiences of families with children with cystic fibrosis in the far north of Brazil", *International Journal of Development Research*, 11, (11), 52183-52185.

## INTRODUCTION

Cystic fibrosis or mucoviscidosis is an autosomal recessive genetic disorder characterized by dysfunction of the *cystic fibrosis transmembrane conductance regulator* (CFTR) gene, which encodes a chlorine transmembrane conductance regulator protein.

It is a complex disease with a progressive and potentially lethal nature, it's changes in the respiratory system and gastrointestinal involvement being the main causes of morbidity and mortality (Feiten, 2018; Raskin et al., 2008; Oliveira et al., 2017).

In the most severe cases of the disease, the obstruction of the pancreatic ducts by mucus starts during intrauterine life, causing pancreatic insufficiency at birth or shortly after birth. The obstruction of the pancreatic ducts impairs the release of enzymes into the duodenum, which causes poor digestion and malabsorption of fats, proteins and carbohydrates, frequently causing chronic diarrhea, with bulky and fatty stools and protein-calorie malnutrition (Silva *et al.*, 2018). Cystic fibrosis is suspected due to changes in the Pezinho Test, a newborn screening test performed between the 3rd and 5th days of age. The findings of two altered tests for cystic fibrosis requires performing the Sweat Test, and two altered results are needed to confirm the diagnosis of cystic fibrosis (Oliveira *et al.*, 2017). The treatment for Cystic Fibrosis is centered around maintaining clear lungs, a good nutritional status - through the supplementation of nutrients and pancreatic enzymes - and antibiotic therapy if infections are present (Pizzignacco *et al.*, 2011). The occurrence of Cystic Fibrosis in children is an intense and complex event, generating a great emotional impact for the family, changes in the family's daily life, plus financial distress, related to the expenses with continuous treatment. Given this approach, this study aims to understand how families are restructured throughout the challenges caused by cystic fibrosis.

## MATERIAL AND METHODS

This is a descriptive cross-sectional study with a qualitative approach, seeking to delve into the subjective aspects and meanings that families attribute to their experiences (Denzin, Lincoln, 2011). Symbolic Interactionism was adopted as the theoretical framework for conducting this study and Narrative Research made it possible to understand the experience “in a process of collaboration between researcher and researched subjects.” (Blumer, 1969; Clandinin, Connelly, 2000). In compliance with Resolution 466/2012 of the National Health Council, the study was approved by the Ethics Committee for Research with Humans, under Opinion No. 4.020.487, CAAE 27848519.10000.0003 (Brasil, 2012). The research was carried out with seven families of eight children diagnosed with cystic fibrosis. The entire universe of children aged between 1 year and six months to twelve years, diagnosed with cystic fibrosis in the state of Amapá until May 2021, was used. The technique used to collect the information was the semi-structured interview, recorded in audio, carried out from March to May 2021. The interviews were carried out with the consent of the participants, after signing the free and informed consent form. To guide the interview, the following proposition was enunciated, with the aim of triggering a narrative, so that the families could report the beginning of the trajectory, following a temporal order regarding the subsequent outcomes: From the diagnosis of Cystic Fibrosis in your child, tell me how it all happened. To ensure confidentiality of information and anonymity, the statements were recorded by degree of kinship and the Letter F, representing the order in which the family was interviewed. The participants' narratives were transcribed in full and coded line by line, as an analytical process, in which concepts were identified and developed in relation to their properties and dimensions (Strauss, Corbin, 1990). Thematic analysis enabled the identification, analysis and reporting of patterns within the data (Holloway, Todres, 2003). The thematic analysis of the narratives resulted in the identification of two themes: The family and the child living with a new routine and Resilience in the trajectory of families.

## RESULTS AND DISCUSSION

**The family and child living with a new routine:** Cystic fibrosis demands intense care, changing the family rhythm and causing physical overload and emotional strain on the family nucleus. “The routine changes a lot, the treatment of cystic fibrosis is intense. It influences study time, bedtime. Requires physical therapy, medications, supplements. She was nine years old, she got worn out” (Mother, F1).

“We had to adapt and reinvent, to take care of a daughter with cystic fibrosis. It's a series of cares, which I try to adapt to her day, to our routine too, so that this is normal for me, and comfortable for her too”. (Mother, F2)

“My life has changed, as I cannot work away from home, as I have to dedicate myself fully to my son. My daily routine is focused around giving medications”. (Mother, F4)

“As it is a chronic disease, our life is focused on getting the treatment right and the necessary follow-up”. (Mother, F7).

Changing family life requires discipline and perseverance for the entire family. The search for ways to adapt appears in an attempt to overcome the difficulties arising from continuous treatment. With regard to the treatment of CF symptoms, Amaral and Rego (2020) highlight the evolution in recent decades, with emphasis on nutritional supplementation, replacement of pancreatic enzymes and daily respiratory physiotherapy associated with previous nebulization with mucolytics, in addition to the recurrent use of antibiotics and anti-inflammatory drugs.

**The resilience in the families' trajectory:** Living with cystic fibrosis represents a challenging experience for the child and their family, however, time is an important ally for acceptance and coping with difficulties.

“There is a moment when the information arrives and you reprocess it, then you suffer and then react”. (Mother, F1).

“Every difficulty overcome constitutes a victory. If I don't face the misfortunes caused by my daughter's illness, everything will be more difficult. So we take it one day at a time. Sometimes wonderful days, other very dark days, but I press on having hope”. (Mother, F3)

“Thinking about the difficulties we've already experienced, cystic fibrosis means overcoming it all. She helped me to become a stronger person and increasingly able to help my son”. (Mother, F4)

“Everything is very difficult, in many situations, despair comes. I thought about dropping everything, but the family found the strength to overcome and follow all the treatment and follow-up”. (Aunt, F5)  
“We went through several difficulties, but we managed”. (Mother, F7)

From the narratives, it is clear that the crises and difficulties caused by CF promoted the development of skills that favored family resilience. In this perspective, Paixão (2020), studying parental control in contexts affected by the illness of children with cystic fibrosis, argues that advances in the use of problem-solving skills and the feeling of self-control can be explained by the strengthening effects, when the family perceives that they have more control over the problems that used to cause them suffering. In the same direction, Santos *et al.* (2017), emphasizes the role of the family nucleus as fundamental, providing protection for its members, in addition to serving as support in coping with the difficulties arising from the chronic disease and its treatment.

## CONCLUSION

From the results presented, it was possible to understand how families are restructured through the challenges caused by cystic fibrosis. The study revealed that what distinguishes resilient families is not the absence of problems, but the ability to face them and the perseverance to overcome them. In the face of these circumstances, we no longer see families as victims and come to understand what they are like because of the way they face adversity. From the narratives, it is clear that the crises and difficulties in life caused by cystic fibrosis promoted the development of skills that favored family resilience.

**Conflict of interests:** The authors declare that the research was carried out in the absence of any commercial or financial relationships that could be interpreted as a potential conflict of interest.

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