ORIGINAL ARTICLE

Characterization of Portuguese Pediatric Patients with Cystic Fibrosis and Their Parents: Cross-Sectional Study

Catarina Guimarães[1], Ana Silva Alexandrino[1], João Antunes Sarmento[2], Inês Azevedo[3]

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Abstract

Introduction: Cystic fibrosis is a chronic disease with multiple associated challenges, not only for the children/adolescents but also for their parents/caregivers. To the best of the authors' knowledge, to this date there are no published studies characterizing the Portuguese pediatric population with cystic fibrosis. This study aims to characterize children/adolescents with cystic fibrosis concerning physiotherapy, hospitalizations in the previous year as well as medication taken in the previous two weeks. We also aimed to characterize the parents/caregivers concerning sociodemographic data, caregivers role (sleep habits, service, and work activities) and equipment sterilization habits for medication administration.

Methods: Cross-sectional study with a total sample of 28 parents/caregivers and 28 children/adolescents (n = 28): 14 were females with an average age of 12.14 ± 3.99 years old. The online question forms - sample selection and characterization questionnaire and care-related quality of life - were sent to the parents/caregivers, through the Associação Portuguesa de Fibrose Quística database.

Results: From the children/adolescents 69.23% (n = 18) had physiotherapy once or twice a week. Most parents/caregivers sterilized the equipment used for medication admission (53.57%) but 33.33% considered that soap and water washing was an effective sterilization.

Discussion: We found that children/adolescents with cystic fibrosis do not undergo physiotherapy at the recommended frequency. In addition, most parents/caregivers have appropriate sterilization habits, although some of them do not have the correct information on how to perform effective sterilization.

Keywords: Adolescent; Child; Cystic Fibrosis/rehabilitation; Health Knowledge, Attitudes, Practice; Parent-Child Relations; Portugal; Surveys and Questionnaires

Introduction

Cystic fibrosis is a hereditary genetic disease with autosomal recessive transmission, which is caused by mutations in the gene encoding the cystic fibrosis transmembrane regulator (*CFTR*), a protein present on the apical surface of epithelial cells. It is classified as a systemic disorder that affects all organs expressing *CFTR*.[1] To date, 2,088 genetic mutations have been identified, which helps to explain the different phenotypes found between affected individuals.[2] The incidence of cystic fibrosis is higher in the Caucasian population and, in Portugal, the *F508del* mutation is the most frequent (46%).[3]

In Portugal, the annual incidence of cystic fibrosis is estimated to be one in 6,000 live births, with an average life expectancy between 35 and 40 years.[4] In recent years, there has been an increase in the survival of these patients justified by the multiple advances in health care, namely the early diagnosis and the evolution of antibiotic therapy. On the other hand, the valorization of pulmonary rehabilitation has contributed not only to increasing the survival of these patients, but mostly to clearly improving their quality of life.[5]

Cystic fibrosis is a complex pathology, demanding a complex management not only from patients but also from their parents/caregivers,[6] with changes in their daily lives being required to ensure correct therapy compliance. These lifestyle changes are most often related to the professional activity, namely resigning from their job, increased number of absences as well as decreased work productivity. As a secondary result, parents may have limitations in social life and

1. Department of Physiotherapy, Escola Superior de Saúde, Instituto Politécnico do Porto, Porto, Portugal
2. Pediatric Cardiology Department, Centro Hospital Universitário de São João, Porto, Portugal
3. Department of Obstetrics-Gynecology and Pediatrics, Faculdade de Medicina, Universidade do Porto, Porto, Portugal
3. Pediatrics Department, Hospital Pediátrico Integrado, Centro Hospitalar Universitário de São João, Porto, Portugal
Corresponding Author
Catarina Guimarães
https://orcid.org/0000-0001-6262-3738
catarina.fidalgoguimaraes@gmail.com
Escola Superior de Saúde, Rua Dr. António Bernardino de Almeida, 400, 4200-072 Porto, Portugal
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recreational activities,[7] culminating in a negative impact of illness and poor perception of happiness.[8] It should be noted that all studies that assess the impact of the disease on the daily life of parents/caregivers as well as the perception of happiness, refer to foreign data, and there is still no concrete data for the Portuguese population.

The objectives of the present study are to:

- Characterize the children/adolescents regarding the current, neonatal, and disease data;
- Characterize the physiotherapy treatments;
- Characterize the medication, hospitalizations during the previous year, and equipment sterilization habits;
- Characterize the caregivers;
- Characterize the disease impact, the happiness perceived by parents/caregivers, and the relationship between both variables.

Methods

Study design

Cross-sectional study according to the Strobe guidelines.[8] The study sample consisted of children/adolescents diagnosed with cystic fibrosis from the Associação Nacional de Fibrose Quística database. The method of diagnosis of cystic fibrosis was based on the guidelines of the Portuguese health authorites (Direção Geral da Saúde), namely neonatal screening, the sweat test and genetic testing. In the present study, an online questionnaire was used, aiming to characterize the sample of children with cystic fibrosis and their respective parents, addressing, for example, the medication used, physiotherapy, and equipment sterilization habits. Since it was an online questionnaire, the authors found it would be easier for caregivers to accurately address the ongoing medication by dividing it into pharmacological groups rather than individualizing each therapy. Sterilization was considered effective when performed for all the equipment after each use.

Participants

The study sample consisted of children/adolescents diagnosed with cystic fibrosis. Selection was made from the Associação Nacional de Fibrose Quística database, and participants were invited to participate through an email sent by association to its members.

The study included children/adolescents aged 6 to 18 years old with a medical diagnosis of cystic fibrosis.[10] The exclusion criteria were children/adolescents with neuro-musculoskeletal disorders and/or neurological deficits as well as history of recent orthopedic surgery (less than six months), since not only could these influence present physical activity levels, but also how to foster a change in the daily parent/caregiver routines. The exclusion criteria for the parents/caregivers were depressive or neurological disorders, and the presence of oncological diseases.

The sample size was calculated using Raosoft® software. An error margin of 20% was considered with a 50% response distribution. A confidence level of 95% was considered and the 150 children/adolescents, members Associação Nacional de Fibrose Quística, were assumed to be the sample size, resulting in a recommended number of participants of 21 children/adolescents.

Therefore, 41 parents answered the questionnaire, 13 of who were excluded for inconsistent health at every size (HAES) response (n = 11) and age over 18 years (n = 2). The final sample comprised 28 children (n = 14 female).

Evaluation tools

Sample selection and characterization questionnaire

To perform the selection and characterization of the sample, the sample selection and characterization questionnaire was prepared. This questionnaire was organized into two sections:

- The first one with a focus on the characterization of the parents/caregivers, addressing caregiving, the impact of cystic fibrosis on the family environment and sleep habits, and other variables. The results regarding the role of the caregiver relate to whoever completed the questionnaire - mother or father - and they were analyzed together.
- The second section was directed to the characterization of the children/adolescents with cystic fibrosis, including some questions about medication, physiotherapy, and equipment sterilization.

Care-related quality of life (CarerQol)

The CarerQol questionnaire, validated for the Portuguese population through the Actifcare project,[11] was designed to assess the impact of a chronic disease on the daily lives of parents/caregivers.[12] This is described as having good content validity and internal consistency.[13] The original work demonstrates good viability, as 98% of the participants completed the questionnaire. The CarerQol questionnaire consists of two parts: CarerQol 7-D and CarerQol VAS.[14]

The CarerQol 7-D consists of seven dimensions that describe the caregiver burden. This includes five negative dimensions: relational problems, mental health problems, problems in combining daily activities, financial problems, and physical health problems. On the other hand, it has two positive dimensions: care

delivery and social support. Caregivers characterized the problems presented in these dimensions using three possible answers: no, some, very.[15]

The answers in the negative dimension range from 0 (very), 1 (some), and 2 (not), and in the positive dimensions range from 0 (not), 1 (some), and 2 (very).[12] The final rating ranges from 0, representing the worst caregiver situation (many problems and no family support/money - greatest negative impact of the disease) to 14, representing the best caregiver situation - lowest negative impact of the disease.[15]

The CarerQol VAS consists of the visual analog scale (VAS), which aims to measure the happiness of the caregiver. The VAS is a horizontal line, where the caregiver indicates a number representing their current happiness, ranging from completely unhappy (0) to completely happy (10), with numerical anchors equally spaced between these two levels.[15]

Variables

The current characterization of children/adolescents: gender (female/male), age (years), current height (m), and current weight (kg).

Neonatal characterization: weeks of gestation, length at birth (cm), weight at birth (kg).

Disease data: age at diagnosis (years), time since diagnosis (years) and diagnostic tests.

Physiotherapy: physiotherapy treatments (yes/no), treatment frequency (1-2 per week, 3-4 per week, > 5 per week), treatment location (physical medicine and rehabilitation clinics, residence, hospital, school) and type of treatment (bronchial clearance, muscle strengthening exercises, aerobic training, stretching, have no information).

Medication: bronchodilators (BD), inhaled antibiotics, oral antibiotics, vitamin supplements (VS), anti-inflammatory (AI), inhaled mucolytics, oral mucolytics, do not take any of these.

Reasons for hospitalizations in the previous year: respiratory status, poor nutritional status, hemoptysis, not related to cystic fibrosis.

Equipment sterilization habits: equipment sterilization (yes/no), instruments that are subject to sterilization (nebulizer masks, nebulizer, tubes, use of disposable material), sterilization method (wash wish soap and water, wash with boiling water, sterilization machine, home care team responsibility), sterilization frequency (after each use, once per day, once per week, two times per week, two times per week and three times per week), information about effective sterilization (doctor, nurse, home care team, physiotherapist).

Caregiver aspect: stopped working to take on the role of caregiver (yes/no), how many days per week do you play the role of caregiver (4-5 days, every day), how many hours a day are you the caregiver (4-8 hours per day, > 8 hours per day), how many hours do you sleep per night (< 5 hours, 6-7 hours, 8 hours), do you have third party support whenever needed (yes/no), who helps you (grandparents, uncles, friends), do you feel you have time for yourself (yes/no), smoking habits (yes/no), smoking habits during pregnancy (yes/no).

Impact of the disease: evaluated by CarerQol 7-D final classification.[15]

Happiness: evaluated using CarerQol VAS.[15]

Statistical analysis

Statistical analysis was performed using the Statistical Package for Social Sciences (SPSS)® software (IBM Corporation, Armonk NY, United States) version 25.0, considering a significance level of 0.05 throughout the analysis.[16] Descriptive statistics was used to characterize the categorical and dichotomous variables through relative frequencies (%), mean and standard deviation.[16] Normality was tested using the Shapiro-Wilk test (n < 30) on the variables related to the current happiness of parents/caregivers and the impact of the disease. Since it was observed that they did not assume a normal distribution, nonparametric tests were used for statistical analysis. Median values, 25th and 75th percentiles (P) were used for descriptive statistics.[16]

Spearman *rho* correlation (rs) was used to relate the impact of the disease on parents/caregivers with the happiness perceived by them.[16]

Results

Characterization of children/adolescents: current, neonatal, and cystic fibrosis characterization

The final sample consisted of 28 children/adolescents (n = 28) aged 6-18 years-old, with a mean age of 12.14 ± 3.99 years. From the children/adolescents, 50% were females. Regarding the characterization of the sample at the neonatal period, the average gestation was 37.46 ± 1.99 weeks. The children were born on average with 3.05 ± 0.40 kg. Eight (28.57%) were premature, only two (7.1%) had a low birth weight (weight < 2500 g). It was found that 5 full-term newborns (17.9%) were in the 50th percentile of weight or higher and seven (25%) were between the 10th and 25th percentiles.

Regarding cystic fibrosis, on average, children were diagnosed at 1.64 ± 0.78 years and twelve were diagnosed in the first month of life. The remaining data regarding the sample characterization can be found in Table 1.

Physiotherapy

It was found that 93% of the sample (n = 26) underwent physiotherapy, and, of those, 69.23% underwent physiotherapy once or twice per week, most frequently at physical medicine and rehabilitation clinics (50%). It was observed that 38.46% of the children underwent a diversified treatment (n = 10) and 42.31% of the parents/caregivers (n = 11) had no information about what was being used as a treatment (Table 2).

Medication, hospitalizations in the previous year, and equipment sterilization habits

When asked about medication, the parents/caregivers mentioned inhaled mucolytics, as the most used (93%). It was found that children under 6 years old (n = 1) were using nebulized hypertonic saline at 3%, and patients over 6 years old were using dornase alfa (92.31%).

It was also found that vitamin supplements were the second most mentioned therapy, either alone (26.9%)

Table 1. Sample characterization in anthropometric measurements: gender, age, weight, and height. Characterization of the neonatal sample: weeks of gestation, length, and weight. Cystic fibrosis characterization: age at diagnosis, time since diagnosis, and complementary means of diagnosis

Variables	Absolute frequency (n)	Mean ± standard deviation	Relative frequency (%)
Gender (%)	14 females 14 males	- -	14 (50%)
Age (years)	28	12.14 ± 3.99	-
Current height (m)	28	1.48 ± 0.22	-
Current weight (kg)	28	38.54 ± 15.97	-
Characterization of the Neonatal and Disease data			
Weeks gestation	28	37.46 ± 1.99	-
Length at birth (cm)	28	47.48 ± 2.30	-
Weight at birth (kg)	28	3.05 ± 0.40	-
Age at diagnosis (years)	28	1.64 ± 0.78	-
Diagnostic time (years)	28	10.79 ± 3.47	-
Complementary diagnostic method			
Sweat test	13	-	46.43%
Neonatal screening + sweat test	15	-	53.57%

Table 2. Physiotherapy: performance, frequency and place of treatment

Variables	Absolute frequency (n)	Relative frequency (%)
Physiotherapy		
Yes	26	93%
No	2	7%
Treatment frequency		
1-2 times per week	18	69.23%
3-4 times per week	5	19.23%
> 5 times per week	3	11.54%
Treatment location		
Physical medicine and rehabilitation clinics	13	50%
Residence	1	3.85%
Hospital	2	7.69%
Residence + physical medicine and rehabilitation clinics	7	26.93%
Residence + hospital + school	3	11.54%
Treatment		
Bronchial clearance	2	7.69%
Muscle strengthening exercises	1	3.85%
Aerobic training	2	7.69%
Stretching	0	-
All previous	10	38.46%
Have no information	11	42.31%

Table 3. Sample characterization regarding the medication of the last two weeks

Variables	Absolute frequency (n)	Relative frequency (%)
Medication		
Bronchodilators	1	3.8%
Inhaled antibiotics	1	3.8%
Oral antibiotics	1	3.8%
Vitamin supplements	7	26.9%
Anti-inflammatory	4	15.4%
Inhaled mucolytics	26	93%
VS+ AI	2	7.7%
VS + BD	4	15.4%
VS + antibiotics + BD + AI	3	11.5%
Do not take any of these	3	11.5%
Hospitalizations in the previous year		
Respiratory exacerbation	5	33.35%
Poor nutritional status	1	6.67%
Respiratory exacerbation + poor nutritional status	1	6.67%
Hemoptysis	1	6.67%
Not related to cystic fibrosis	7	46.67%
Equipment sterilization		
Yes	15	53.57%
No	13	46.43%
Instruments that are subject to sterilization		
Nebulizer masks	10	66.67%
Nebulizer + nebulizer masks	2	13.33%
Nebulizer + nebulizer masks + tubes	2	13.33%
Use of disposable material	1	6.67%
Sterilization method		
Wash with soap and water	5	33.33%
Wash with boiling water	5	33.33%
Wash with soap and water + boiling water	2	13.33%
Sterilization machine	1	6.67%
Home care team responsibility	1	6.67%
Boiling water wash + home care team responsibility	1	6.67%
Sterilization frequency		
After each use	6	40%
Once per day	2	13.33%
Once per week	4	26.66%
Twice per week	2	13.33%
Three times per week	1	6.67%
Information for effective sterilization		
Doctor	7	46.67%
Nurse	3	20%
Home care team	2	13.33%
Physiotherapist	0	-
Doctor + home care team + nurse	2	13.33%
All of the above	1	6.67%

AI - anti-inflammatory Drugs; BD - bronchodilators; VS - vitamin supplements.

or in association with other medications like anti-inflammatory drugs (7.7%) and bronchodilators (15.4%). Furthermore, 53.6% (n = 15) of the children were hospitalized in the previous year, and 86.7% (n = 13) of these patients were admitted to the hospital once. The main reason for hospitalization, when related to cystic fibrosis, was the worsening of the patients' respiratory condition (33.37%).
Regarding the sterilization of medical equipment, from the 53.57% (n = 15) who reported sterilizing the equipment, 67% were only sterilizing the masks used for nebulization. Most parents/caregivers received the information for effective sterilization from the attending physician (46.67%) (Table 3).

Caregiver aspect

Therefore, 96.43% of the parents/caregivers provided care every day, with 39.29% (n = 11) having to quit their jobs. When asked about sleeping habits, most of the parents/caregivers reported sleeping between 6 and 7 hours per night (60.71%) (Table 4).

Disease impact, happiness perceived by the parents/caregivers, and the relationship between both

Regarding the happiness perceived by the parents/caregivers, 25% of the parents/caregivers (n = 7) considered that their current happiness was 8 out of 10 possible, and 28.6% (n = 8) considered it to be 9 out of 10. Regarding the impact of the disease on the parents/caregivers, 25% (n = 8) of the parents/caregivers had an average impact of the disease - 8 out of 14 points possible - and 10.7% had a final result of 9 out of 14 possible.
The impact of the disease on the parents/caregivers was related to the current happiness perceived by

Table 4. Characterization of the parents/caregivers in care delivery

Variables	Absolute frequency (n)	Relative frequency (%)
Have you stopped working to take on the role of caregiver?		
Yes	11	39.29%
No	17	60.71%
How many days per week do you play the role of caregiver?		
4-5 days	1	3.57%
Every day	27	96.43%
How many hours a day are you the caregiver?		
4-8 hours per day	13	46.43%
> 8 hours per day	15	53.57%
How many hours do you sleep per night?		
< 5 hours	4	14.29%
6-7 hours	17	60.71%
8 hours	7	25%
Do you have third party support whenever needed?		
Yes	18	64.29%
No	10	35.71%
Who helps you?		
Grandparents	14	77.78%
Grandparents and uncles	2	11.1%
Uncles	1	3.57%
Friends	1	3.57%
Do you feel you have time for yourself?		
Yes	14	50%
No	14	50%
Smoking habits		
Yes	17	60.71%
No	11	39.29%
During the pregnancy		
Yes	5	17.86%
No	23	82.14%

them, with a strong positive relationship between these two (rs = 0.734). It was found that the lower the negative impact of the disease on the life of the parents/caregivers is, the greater the happiness was that they perceived (p < 0.001), with this relationship being bidirectional (Table 5).

Table 5. Relationship between the impact on the caregiver and the happiness perceived by them

Variables	Median (P25, P75)	rs	*p*
Caregivers impact	8 (6; 9)	0.734	< 0.001
Happiness	8 (7; 9)		

P25 - 25th percentile; P75 - 75th percentile; rs - Spearman's correlation.

Discussion

Through the analysis of the sample of the children/adolescents, it was observed that the diagnosis of cystic fibrosis was made on average during the first two years of life. Due to early diagnosis, the average life expectancy and perceived quality of life of the patients has increased as they begin early appropriate medical therapy and physiotherapy.[17] For this reason, there has been a decrease in the number of hospitalizations over the years.[18] Nevertheless, a large part of the target study sample was hospitalized during the previous year, having as a main cause the worsening of the respiratory status related to cystic fibrosis. The higher number of hospitalizations observed may be due to non-compliance with the prescribed therapy either alone or in association with other types of medication.[19] Studies report that cystic fibrosis patients adhere more easily and comply with therapy when it has immediate effects compared to those that have a medium/long term effect.[20] On the other hand, the same study states that adolescents with cystic fibrosis have less knowledge of their daily therapy compared to their parents/caregivers, indicating that they are not prepared to take control or make decisions about therapy when they reach adulthood.

In this sample, most children attended physiotherapy treatments, predominantly in physical medicine and rehabilitation clinics, hospitals, or at home. Through analysis of the data, it was demonstrated that the patients of the sample had a complete treatment - aerobic training, muscle strengthening, flexibility, and bronchial hygiene - in line with the international guidelines, which establish that the treatment should not only be directed to bronchial hygiene, but also with an emphasis on the cardiovascular component (exercise and aerobic training).[21] Although it was observed that the treatment plan followed international recommendations, most of the children/adolescents only undergo physiotherapy once or twice per week, which is lower than recommended in the international cystic fibrosis physiotherapy guidelines, where daily or twice a day physiotherapy is recommended.[22]

It is noteworthy in this study that there is a lack of information from the parents/caregivers about the treatment that the children/adolescents perform, which may be an indicator of the poor empowerment of parents/caregivers for the importance of physiotherapy treatments.[21] Previous studies suggest that those parents/caregivers who know how to correctly perform treatments and how to recognize symptoms suggestive of cystic fibrosis exacerbation can have an impact by minimizing the burden of exacerbations.[23]

Regarding the characterization of the parents/caregivers, a large part quit their jobs to assume the role of caregiver. At the same time, most of the parents/caregivers in this study sleep an average of 6-7 hours per night. Thus, the parents/caregivers follow the trend observed in other studies regarding the parents of children with chronic diseases,[24] being below the 8-9 hours of sleep recommended in the literature.[25]

On the other hand, sleep deprivation contributes to the parents/caregivers becoming increasingly exhausted, affecting care delivery, daily living tasks, and the family relationship itself.[24] The parent/caregiver burnout may not only be related to the aforementioned aspects but also to the lack of care assistance.[26] In the present study, most of the parents/caregivers had third-party support in providing care, with grandparents being the most common help. For a better characterization of the aid by third parties, further studies will be needed.

Regarding equipment sterilization habits, most of the parents/caregivers claim to have these habits. There is a clear predominance in the sterilization of masks used for aerosol therapy, probably related to the type of medication prescribed, namely bronchodilators and inhaled antibiotics for the management of respiratory infections.[19] On the other hand, some of the parents/caregivers consider that they sterilize the equipment only by washing it with soap and water. This may correlate to a lack of knowledge about physiotherapy treatments and failure to provide adequate instructions. Education sessions may be missing for caregivers of cystic fibrosis with daily life issues.[27] It should also be noted that none of the parents/caregivers received instructions for effective sterilization from physiotherapists, which is an important factor because this professional class is

present in a large part of the treatment (in this study, once or twice per week).

A limitation of the present study is the fact that the completion of the questionnaires was online, making it more difficult to understand, with consequent ambiguities in the responses. Furthermore, the study is cross-sectional, using a convenience sample, with its inherent limitations. Moreover, the unsupervised filling out of a questionnaire could bring a measurement bias to the study, as some caregivers may have not understood the questions or may have not answered them accurately. In future studies, better information and instructions will be required for participants, reducing the possible existing measurement bias.[28]

In fact, most patients with cystic fibrosis have pancreatic insufficiency and, the patient nutritional status, can be related to lung function. The present study did not evaluate the intake of pancreatic enzymes, nor the present nutritional status in detail, aspects that should be evaluated in future studies. Finally, the authors did not discriminate each individual medication prescribed, rather placing them into pharmacological groups. While it may have helped the caregivers to fill the questionnaire, it may have had an impact on the conclusions about therapy compliance.

It was found that the children/adolescents with cystic fibrosis in our population do not undergo physiotherapy at the recommended frequency but have widespread access to treatments, with an emphasis on aerobic training and bronchial clearance.

Most patients of the sample were hospitalized during the previous year, having as the main cause the worsening of the respiratory condition related to cystic fibrosis. On the other hand, vitamin supplements were the most used therapy reported by parents, either alone or in conjunction with other types of medication - bronchodilators, antibiotics, and anti-inflammatory drugs.

Regarding the parents/caregivers, it was found that they have less than ideal sleeping habits regarding what is recommended in the literature. Most of the parents/caregivers provide care every day, and most of them have permanently left work.

The parents/caregivers have a medium impact from the disease and at the same time a good perception of their current happiness. The impact of the disease and their perceived happiness are closely related.

Finally, it was found that most of the parents/caregivers claim to have sterilization habits, with a large part only sterilizing the masks used for nebulization and washing the equipment with soap and water. The authors also hypothesize that some of the parents/caregivers may have not been provided with adequate information on how to perform effective sterilization.

WHAT THIS STUDY ADDS

- To the best of our knowledge, there are no published studies characterizing the Portuguese pediatric population with cystic fibrosis and their parents.
- The characterization of the physiotherapy sessions was carried out, describing the positive and negative aspects found.
- The present study aimed to simultaneously characterize the parents of the children with cystic fibrosis and to analyze the impact of the disease on the family.
- This study emphasizes the need for clarification sessions on various topics, such as correct equipment sterilization by parents.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Provenance and peer review

Not commissioned; externally peer reviewed

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Caracterização dos Doentes Pediátricos Portugueses com Fibrose Quística e dos Seus Pais: Estudo Transversal

Resumo:

Introdução: A fibrose quística é uma doença crónica que tem múltiplos desafios associados, não apenas para as crianças/adolescentes, como também para os seus pais/cuidadores. Tanto quanto é do conhecimento dos autores, não foram publicados até à data estudos que caracterizem a população pediátrica portuguesa com fibrose quística. O presente estudo teve como objetivo caracterizar crianças/adolescentes com fibrose quística quanto à fisioterapia, internamentos no ano anterior, bem como medicação nas duas semanas anteriores. Caracterizaram-se também os pais/cuidadores quanto aos dados sociodemográficos, papel do cuidador (hábitos de sono, serviço e atividade laboral) e hábitos de esterilização de equipamentos de administração de medicamentos.

Métodos: Estudo transversal com uma amostra de 28 pais/cuidadores e 28 crianças/adolescentes (n = 28): 14 do sexo feminino com uma idade média de 12,14 ± 3,99 anos. Os formulários eletrónicos - questionário de seleção e caracterização da amostra e qualidade de vida relacionada com os cuidados - foram enviados aos pais/cuidadores, através da base de dados da Associação Portuguesa de Fibrose Quística.

Resultados: Das crianças/adolescentes, 69,23% (n = 18) fizeram fisioterapia uma ou duas vezes por semana. A maioria dos pais/cuidadores esterilizou o equipamento utilizado na administração da medicação (53,57%), mas 33,33% considerou que a lavagem com água e sabão era um método de esterilização eficaz.

Discussão: As crianças/adolescentes com fibrose quística não realizam fisioterapia na frequência recomendada. Além disso, a maioria dos pais/cuidadores tem hábitos de esterilização adequados, embora alguns não estejam na posse de informações corretas sobre como realizar uma esterilização eficaz.

Palavras-Chave: Adolescente; Conhecimentos, Atitudes e Prática em Saúde; Criança; Fibrose Quística/reabilitação; Inquéritos e Questionários; Portugal; Relações Pais-Filho