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ABSTRACT

Introduction: Cystic fibrosis (CF) is a chronic genetic disease with multisystemic impact, primarily respiratory, which can compromise functioning in children and adolescents. The Pediatric Evaluation of Disability Inventory – Computer Adaptive Test (PEDI-CAT) is a functional tool based on the International Classification of Functioning, Disability and Health (ICF), with potential to assess multiple domains of functioning. **Objective:** To evaluate the relationship between functioning, clinical severity, and respiratory impairment in children and adolescents with CF. **Material and Methods:** A cross-sectional observational study was conducted with 26 participants aged 6 to 21 years with a confirmed diagnosis of CF. Anthropometric and spirometry characteristics (forced expiratory volume – FEV1), functional capacity (Six-Minute Walk Test – 6MWT), Shwachman-Kulczycki score, and functioning (PEDI-CAT) were assessed. Correlations were analyzed using Pearson’s coefficient, and group comparisons by t-test. **Results:** All PEDI-CAT domains showed a positive and moderate correlation with FEV1 ($r = 0.46-0.57$; $p < 0.05$), and the mobility domain was positively and moderately correlated with clinical severity ($r = 0.48$; $p = 0.01$). No significant correlations were observed between PEDI-CAT domains and the 6MWT. Children with greater respiratory obstruction presented higher scores in the responsibility domain of the PEDI-CAT ($p < 0.05$). **Conclusion:** Functioning, as measured by the PEDI-CAT, is related to clinical and respiratory severity in CF. The instrument proved to be sensitive in discriminating functional differences, particularly in the responsibility domain.

Keywords: Cystic Fibrosis; Functional Status; Child; Adolescent; Respiratory Function Tests.

RESUMO

Introdução: A fibrose cística (FC) é uma doença genética crônica de impacto multissistêmico, predominantemente respiratório, que pode comprometer a funcionalidade de crianças e adolescentes. O Pediatric Evaluation of Disability Inventory – Computer Adaptive Test (PEDI-CAT) é um instrumento funcional baseado na Classificação Internacional de Funcionalidade, Incapacidade e Saúde (CIF), com potencial para avaliar múltiplos domínios da funcionalidade. **Objetivo:** Avaliar a relação entre a funcionalidade, a gravidade clínica e o comprometimento respiratório em crianças e adolescentes com FC. **Material e Métodos:** Estudo observacional transversal, realizado com 26 participantes, com idades entre 6 e 21 anos, com diagnóstico confirmado de FC. Foram avaliadas as características antropométricas e espirométricas (volume expiratório forçado no primeiro segundo – VEF1), a capacidade funcional (Teste de Caminhada de Seis Minutos – TC6), o escore de Shwachman-Kulczycki e a funcionalidade por meio do PEDI-CAT. As correlações foram analisadas pelo coeficiente de Pearson, e as comparações entre grupos, pelo teste t. **Resultados:** Todos os domínios do PEDI-CAT apresentaram correlação positiva e moderada com o VEF1 ($r = 0,46-0,57$; $p < 0,05$), e o domínio mobilidade mostrou correlação positiva e moderada com a gravidade clínica ($r = 0,48$; $p = 0,01$). Não foram observadas correlações significativas entre os domínios do PEDI-CAT e o TC6. Crianças com maior obstrução respiratória apresentaram escores mais elevados no domínio responsabilidade do PEDI-CAT ($p < 0,05$). **Conclusão:** A funcionalidade, avaliada pelo PEDI-CAT, relaciona-se com a gravidade clínica e respiratória na FC. O instrumento mostrou-se sensível para discriminar diferenças funcionais, especialmente no domínio responsabilidade.

Palavras-chave: Fibrose cística; Estado funcional; Criança, Adolescente, Testes de função respiratória.

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INTRODUCTION

Cystic fibrosis (CF) is a progressive, multisystemic, autosomal recessive genetic disorder caused by mutations in the *Cystic Fibrosis Transmembrane Conductance Regulator* (CFTR) gene, located on chromosome 7 (7q31.2).^{1,2} These mutations impair the CFTR protein, which regulates ion transport across epithelial surfaces, leading to thick, viscous secretions that primarily affect the lungs and gastrointestinal tract.^{3,4}

A study using data from the Canadian CF Registry and the U.S. Cystic Fibrosis Foundation Patient Registry (CFFPR) from 1995 to 2019 reported a 1.6% annual decrease in incidence, with rates of 1:3,848 in Canada and 1:5,078 in the United States in 2019.⁵ It is estimated that about 162,000 people worldwide live with CF, including approximately 57,000 who remain undiagnosed, with the highest burden observed in India.⁶

Clinical manifestations are wide-ranging and include recurrent respiratory infections, steatorrhea, and malnutrition.¹ Thick mucus in the lungs promotes inflammation, chronic infections, and bronchiectasis. CFTR dysfunction also affects the gastrointestinal tract, causing pancreatic insufficiency, malabsorption, malnutrition, and diabetes. In newborns, signs such as meconium ileus, prolonged jaundice, and salt imbalance may indicate the disease, while older children and adults may present with pancreatitis and male infertility.^{1,7}

Since 2010, Brazil's neonatal screening program has enabled early diagnosis and interventions aimed at preserving lung function and nutritional status, controlling infections, and, in severe cases, considering lung transplantation.^{1,8,9} With the advent of CFTR modulator therapies, survival and quality of life have improved.^{8,10} Nevertheless, respiratory limitations, malnutrition, hospitalizations, and psychosocial impacts continue to interfere with daily activities and school performance.^{11,12}

CF poses significant challenges during childhood and adolescence because of substantial phenotypic variability—even among patients with the same genotype—suggesting the influence of environmental and genetic modifier factors.^{13,14} Treatment requires a multidisciplinary team with clinical, physiotherapeutic, and nutritional focus, along with ongoing monitoring of pulmonary function and functional capacity. However, available assessment tools primarily emphasize respiratory structural aspects and overlook activities and participation, domains highlighted by the International Classification of Functioning, Disability and Health (ICF).¹⁵

Few studies have evaluated functioning based on this model in children and adolescents with CF.¹⁶ Understanding this dimension may help refine therapeutic goals and guide more effective interventions. Therefore, this study aimed to analyze the relationship between functioning, assessed by the Pediatric Evaluation of Disability Inventory – Computer Adaptive Test (PEDI-

CAT), and clinical and respiratory severity, measured by the Shwachman-Kulczycki score and forced expiratory volume in one second (FEV1). Additionally, functional capacity was evaluated using the Six-Minute Walk Test (6MWT), and comparisons were performed across different levels of respiratory severity.

MATERIAL AND METHODS

Design and ethics

This observational, cross-sectional study was conducted in accordance with the STROBE guidelines and the ethical principles of the Declaration of Helsinki. The study protocol was approved by the Research Ethics Committee of the University Hospital of the Universidade Federal de Juiz de Fora (UFJF) (No. 3.299.985). Data were collected between July 1, 2019, and October 20, 2019. Legal guardians provided written informed consent, and participants provided assent when appropriate.

Participants

Individuals aged 6 to 21 years with a confirmed diagnosis of cystic fibrosis and receiving care at the specialized outpatient clinic of the University Hospital–UFJF were included. Exclusion criteria were: respiratory exacerbation within the previous four weeks; neurological or cognitive impairment that prevented test performance; or inability to comprehend the assessment instruments.¹⁷

Samples size calculation

The sample size was estimated using the statistical platform of the University of California, San Francisco (UCSF) (<https://data.ucsf.edu/research/sample-size>) to detect a two-tailed correlation coefficient significantly different from zero. For the study design, a Type I error rate (α) of 0.05 and a Type II error rate (β) of 0.20 were established, ensuring a statistical power of 80%. Based on an expected correlation coefficient (r) of 0.50₇ between the PEDI-CAT and the Six-Minute Walk Test (6MWT), a total sample size of 29 participants was required. The calculation followed the methodological guidelines of Hulley et al,¹⁸ utilizing the Fisher transformation to normalize the data.

Procedures

Researchers underwent prior training to standardize the use of instruments. A screening questionnaire collected demographic and clinical data. Participants then underwent anthropometric assessment, spirometry, and the Six-Minute Walk Test (6MWT) in random order.^{19,20,21} Anthropometric evaluation

followed the recommendations of the International Society for the Advancement of Kinanthropometry (ISAK).¹⁹ Clinical severity was determined using the Shwachman–Kulczycki score from medical records, and functioning was assessed with the Pediatric Evaluation of Disability Inventory – Computer Adaptive Test (PEDI-CAT), administered as a computer-based interview with parents or legal guardians.^{22,23,24} (Figure 1)

Body mass and height

Body mass (kg) and height (m) were measured using an electronic scale and a portable stadiometer, following ISAK recommendations.¹⁹ Participants stood upright with heels together, arms relaxed, and head in the horizontal plane. All measurements were taken by a trained nutritionist. Body mass index (BMI) was calculated as weight divided by height squared.²⁵ Height-for-age and BMI-for-age were determined using the WHO

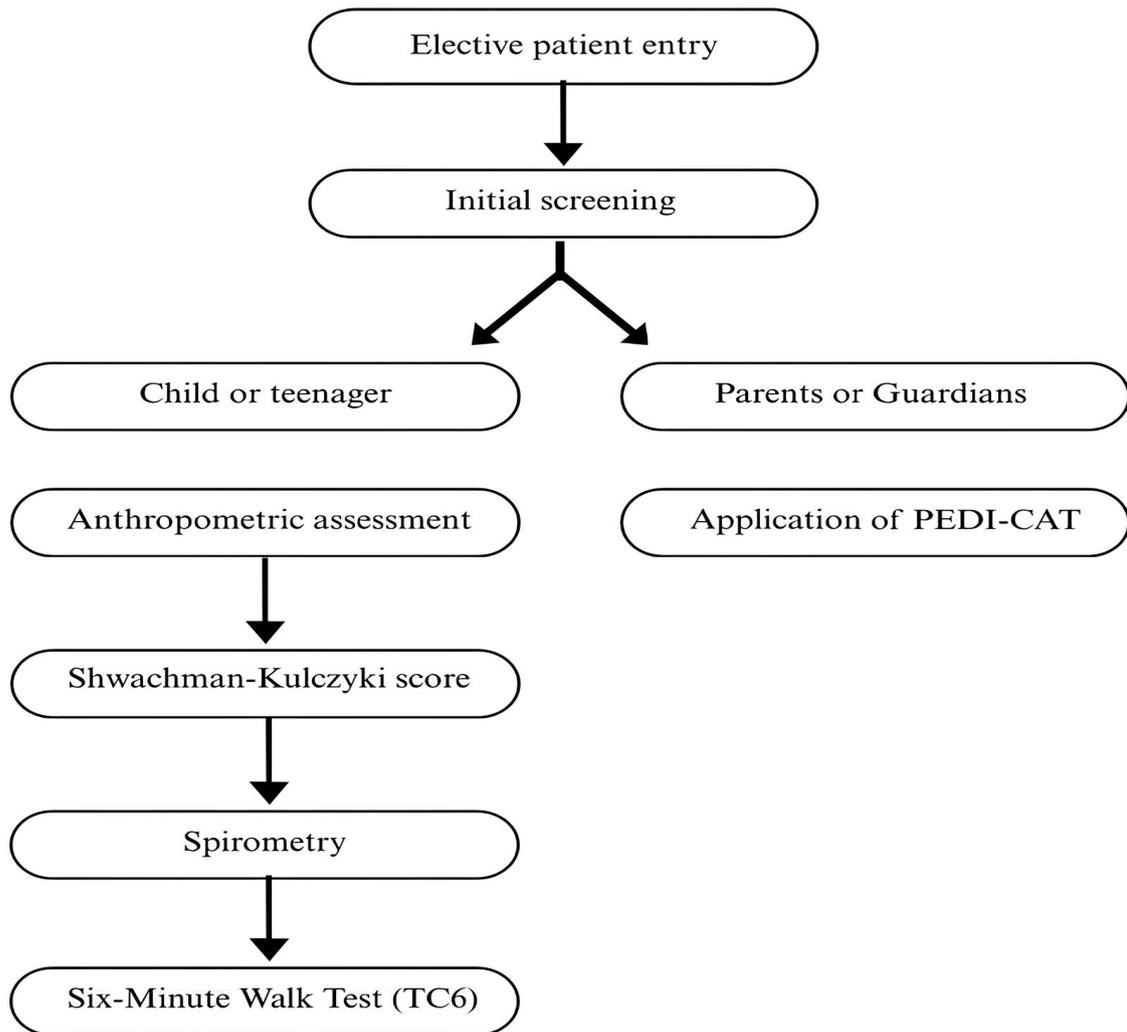


Figure 1. Assessment procedures

Anthro Plus software and classified according to World Health Organization cutoff points.¹⁵

Shwachman-Kulczycki score

The Shwachman–Kulczycki score was used to assess the clinical severity of participants and was completed by the attending physician based on medical records.²² This instrument was developed to measure disease severity and the impact of therapeutic interventions, encompassing four domains: general activity, physical findings, nutritional status, and radiological findings. Each domain is scored in five categories ranging from 5 (worst) to 25 points (best), for a total possible score of 100. The overall score classifies health status into five levels: excellent (86–100 points), good (71–85), mild (56–70), moderate (41–55), and severe (≤ 40).²⁶

Pediatric evaluation of disability inventory computer adaptive test (PEDI-CAT)

The PEDI-CAT is designed to evaluate the functioning of children and adolescents by considering their ability to perform daily activities and participate in various settings.^{23,24} The instrument consists of a bank of 276 items covering four functional domains: (1) Daily Activities, (2) Mobility, (3) Social/Cognitive, and (4) Responsibility. These domains can be administered individually or together, and responses can be collected through interviews with parents or caregivers or by clinical judgment from health professionals familiar with the child's routine.^{23,24}

The PEDI-CAT software uses a computer adaptive testing (CAT) algorithm that selects items based on the participant's age, sex, and previous responses. Each domain begins with the same initial question, and subsequent items become easier or more difficult depending on the given answer. This adaptive process ensures that questions match the child's functional level, reducing the total number of items administered and eliminating irrelevant questions without compromising accuracy. The instrument uses accessible language and, in some domains, illustrations to enhance comprehension.^{23,24}

Two software versions are available: the Speedy Version ("Precise"), which administers 5–15 items per domain, and the Content-Balanced Version ("Comprehensive"), which is longer and recommended for therapeutic planning. For the present study, the Speedy Version was used, which presents greater agility in administration while maintaining equivalent reliability of results presents greater agility in administration while

maintaining equivalent reliability.^{23,24}.

The system provides several scoring formats: age-normative percentiles, T-scores, a continuous score (scale from 20 to 80), and an item-response report.^{23,24} In this study, the continuous score was chosen, allowing comparisons among participants. Within the framework of the International Classification of Functioning, Disability and Health (ICF), the PEDI-CAT domains correspond to the dimensions of activity and participation.

Spirometry

Spirometry was performed on all participants using a Koko® spirometer by a trained professional. The procedure was explained and demonstrated beforehand. Testing was carried out in a seated position with feet and back supported, using a nose clip and holding the mouthpiece with both hands. Each maneuver consisted of a maximal inspiration followed by a forced expiration lasting at least six seconds, with three to eight valid attempts. Technical and reproducibility criteria followed the recommendations of the Brazilian Thoracic Society.²⁰ Measurements included forced vital capacity (FVC), forced expiratory volume in one second (FEV₁), the FEV₁/FVC ratio, and forced expiratory flow between 25% and 75% of FVC (FEF_{25–75%}), all compared with national reference values.²⁷ The primary outcome variable was FEV₁, expressed in absolute values and as a percentage of predicted values, representing respiratory function according to the ICF framework.

Six-minute walk test (6MWT)

The 6MWT is a self-paced exercise capacity test in which participants walk as far as possible for six minutes along a flat corridor at least 30 m in length.²¹ Each participant performed the test twice to minimize learning effects, under the supervision of the same evaluator, with standardized verbal encouragement every minute and during the final 15 seconds. In cases of intense dyspnea or fatigue, participants were instructed to pause without stopping the timer and to resume walking as soon as possible.

Heart rate (Polar monitor), blood pressure, peripheral oxygen saturation (Nonin), and perceived dyspnea and fatigue (modified Borg scale) were recorded before and after the test.²⁸ Distance walked was compared with predicted values for Brazilian children aged 7–12 years using sex-specific equations:

- Boys: Distance (m) = [16.86 × age (years)] + [1.89 × Δ heart rate (beats/min)] – [0.80 × weight (kg)] + (336.91 × R₁)
- Girls: Distance (m) = [13.54 × age (years)] + [1.62 × Δ heart rate (beats/min)] – [1.28 × weight (kg)] + (352.33 × R₁).

The R_1 coefficient was adjusted for the Southeast region of Brazil. The outcome variable was the distance covered (6MWD), expressed in meters and as a percentage of the predicted value, using the best performance obtained.

Statistical analysis

Data normality was assessed using the Shapiro–Wilk test. Continuous variables were expressed as mean \pm standard deviation or as median and interquartile range, depending on distribution, and categorical variables as frequency and proportion. Correlations among functioning (PEDI-CAT), distance covered in the Six-Minute Walk Test (6MWT), FEV₁, and Shwachman–Kulczycki (SK) score were analyzed using Pearson's correlation coefficient, interpreted as very strong ($r > 0.90$), strong (0.70–0.90), moderate (0.50–0.70), weak (0.30–0.50), or negligible (0–0.30).²⁹ Group comparisons across different levels of respiratory severity were performed with an unpaired t test. A significance level of 5% ($\alpha = 0.05$) was adopted. Analyses were conducted using SPSS software, version 20.0.

RESULTS

Participants characteristics

This study was conducted in a specialized cystic fibrosis outpatient clinic. At the time of recruitment, 37 patients were registered at the service. Among them, seven were excluded because they were younger than 6 years old and four were excluded because they were

older than 21 years. Therefore, 26 patients met the eligibility criteria and were all included in the study. A participant flow diagram describing the recruitment process and exclusions was included according to STROBE recommendations.

The mean age was 11.6 ± 4.6 years, with a predominance of males (73%). The mean BMI-for-age z-score was -0.79 ± 1.76 , indicating mild nutritional impairment. Regarding pulmonary function, 14 participants had FEV₁ $> 70\%$ (mild classification), seven were classified as moderate (FEV₁ 41–70%), and one participant showed severe obstruction (FEV₁ $< 40\%$). The mean Shwachman–Kulczycki score was 83.3 ± 17.8 , reflecting mild to moderate clinical severity. Functional capacity assessed by the 6MWT showed that all tested participants ($n = 13$, age < 13 years) walked more than

Table 1: Descriptive characteristics of the participants with Cystic Fibrosis.

Variables		Values
Demographics	Age (years)	11.6 \pm 4.6
	Male, n (%)	19 (73.1)
Anthropometry	BMI Score	-0.79 \pm 1.76
Lung Function	VEF1	72.4 \pm 18.9
	Mild	14 (54%)
	Moderate	7 (27%)
	Severe	1 (4%)
Physical Capacity	6MWT (% predicted)	>80% in all
Clinical Severity	SK Score	83.3 \pm 17.8

Legend: Forced expiratory volume in one second (FEV₁); Six-minute walk test (6MWT); Shwachman-Kulczycki (SK) score; Body mass index (BMI).

80% of the predicted distance (Figure 2), (Table 1).

Correlations between functioning, pulmonary function, and clinical severity

Correlations of PEDI-CAT scores with 6MWT distance, FEV₁, and total Shwachman–Kulczycki scores are presented in Table 2. FEV₁ showed positive, moderate correlations with all PEDI-CAT domains (Daily Activities, Mobility, Social/Cognitive), while the clinical severity score (total SK) demonstrated a positive, moderate correlation only with the Mobility domain. No significant correlations were observed between PEDI-CAT scores and 6MWT distance.

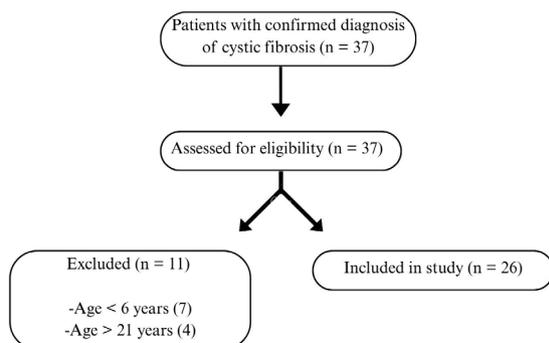


Figure 2. Flowchart of participant selection

Table 2: Correlation between PEDI-CAT domains and the variables 6MWT, Total SK and FEV1

Variables	6MWT		VEF1		TOTAL SK	
	r	p	r	p	r	p
Daily Activities	0.14	0.54	0.56	<0.001*	0.38	0.06
Mobility	0.09	0.68	0.57	<0.001*	0.48	0.01*
Social/Cognitive	-0.01	0.62	0.54	<0.001*	0.19	0.35
Responsibility	-0.01	0.67	0.46	0.030*	0.29	0.15

Legend: Forced expiratory volume in one second (FEV1); Six-minute walk test (6MWT); Total Shwachman-Kulczyki score (SK Total); r: Pearson correlation; PEDI-CAT: Pediatric Evaluation of Disability Inventory Computer Adaptive Test; p<0.05.

Comparasion by respiratory severity

As shown in Table 3, participants with moderate to severe ventilatory obstruction were older, had greater clinical severity, and exhibited higher scores in the responsibility domain of the PEDI-CAT compared with those with mild obstruction.

The PEDI-CAT was easy to administer, quick (approximately 12 minutes), and easily understood by parents or caregivers. Its various domains showed moderate correlations with FEV₁, a marker of ventilatory obstruction and respiratory severity. Although no gold-standard instrument exists for assessing functioning in children and adolescents with CF, functional capacity tests such as the 6-Minute Walk Test (6MWT) are widely

Table 3: Comparison of PEDI-CAT domains according to respiratory impairment severity

Variables	Light	Moderate/severe	p
	(FEV1>60% pred)	(FEV1<60% pred)	
	n=18	n=8	
Age	10.00 ± 3.11	15.38 ± 5.18	0.006*
Total SK	93.57 ± 7.45	67.59 ± 16.69	<0.001*
Daily Activities	66.16 ± 2.91	66.25 ± 3.24	0.957*
Mobility	74.50 ± 2.91	73.75 ± 2.05	0.792*
Social/Cognitive	72.50 ± 3.79	73.25 ± 1.58	0.598*
Responsibility	53.36 ± 8.11	58.75 ± 7.62	0.032*

Legend: Forced expiratory volume in one second (FEV1); predicted percentage (% pred); Shwachman-Kulczyki score (SK); PEDI-CAT: Pediatric Evaluation of Disability Inventory – Computer Adaptive Test. *p < 0.05.

DISCUSSION

To the best of our knowledge, this is the first study to investigate functioning in children and adolescents with cystic fibrosis (CF) using the PEDI-CAT. The results demonstrate that the PEDI-CAT domains are associated with respiratory and clinical severity, while the Responsibility domain showed higher scores among older participants and those with moderate to severe ventilatory obstruction. This finding likely reflects greater autonomy and increased involvement in disease management among adolescents, rather than greater functional impairment in younger children.

used across age groups. Studies using the 6MWT as a marker of functional capacity have also shown moderate correlations with respiratory severity.³⁰ Regarding the total clinical severity score, a moderate correlation was observed only with the mobility domain. Similarly, Ozipek reported worse mobility in patients with greater clinical severity, emphasizing that frequent exacerbations, low aerobic capacity, and respiratory symptoms can impair functioning and quality of life.³¹ Comparable results have been observed in adults with CF, where better functional scores were associated with lower respiratory severity.³²

In contrast, none of the PEDI-CAT domains were associated with 6MWT distance. The 6MWT is widely used in chronic respiratory diseases to assess physical

functional capacity and to predict morbidity, mortality, and response to interventions. However, in children and adolescents with CF, its sensitivity is controversial, as individuals with significantly impaired lung function may still perform satisfactorily on the test.³³ The 6MWT likely has methodological limitations that reduce its effectiveness as a functional marker, particularly when compared with adult respiratory populations. This may be explained by the test's submaximal nature, its self-paced speed, prohibition of running, time limitation, and minimal motivational or playful stimulus for pediatric participants.³⁴

In the present study, all participants who met the age criteria for the predictive equation (≤ 13 years) exceeded 80% of the predicted 6MWT distance, suggesting a ceiling effect in this pediatric population. A systematic review highlighted the limitations of the 6MWT in children with less severe pulmonary impairment due to a ceiling effect related to exercise intensity.³⁰ Therefore, the test may not be sensitive enough to discriminate physical functional capacity in this population, except in severe cases where sarcopenia, physical inactivity, and inflammation contribute to reduced mobility. These findings highlight the need for more sensitive and easily administered assessment tools, with the PEDI-CAT emerging as a promising alternative.

When comparing functioning across different levels of respiratory severity in children and adolescents with CF, those with greater ventilatory obstruction had higher scores in the responsibility domain compared to those with mild disease, with no differences observed in other PEDI-CAT domains. Managing CF requires substantial responsibility, including airway clearance, nebulization, medications, and a specific diet.³⁵ Low adherence can lead to increased infections, faster disease progression, hospitalizations, reduced quality of life, and premature mortality. In the present study, participants with moderate-to-severe disease were significantly older and demonstrated higher Responsibility scores. This finding appears to be primarily influenced by developmental factors and the progressive transfer of self-care tasks throughout adolescence, particularly in the context of greater clinical complexity, rather than by disease severity alone. Murphy reports that parents of children with CF assume significant responsibility for disease management, including adherence monitoring, medication administration, facilitation of respiratory exercises, and emotional support.³⁶ In contrast, young adults with CF are capable of assuming responsibility for self-care, provided they are supported by a structured transition plan, effective communication, and coordination with healthcare professionals.³⁷ These findings support the present study results, in which children with milder CF exhibited lower responsibility than adolescents with moderate-to-severe disease, who demonstrated greater autonomy.

The PEDI-CAT is a multidimensional,

comprehensive, and easily administered instrument capable of evaluating different levels of functioning and disability in children and adolescents with CF. Beyond describing functioning domains, the PEDI-CAT may contribute to a more comprehensive understanding of functional performance in clinical practice. In this way, it provides useful information for researchers and clinicians to plan the progressive transfer of autonomy and self-care, establishing clear rehabilitation goals.²⁴

The study employed a convenience sample, which may limit the generalizability of the findings. Although the estimated sample size was 29 participants, this number was not reached due to the limited availability of eligible patients during the data collection period, as well as participant refusals and operational constraints within the service. Therefore, the results should be interpreted with caution, with emphasis placed on the magnitude of the associations and their corresponding confidence intervals.

CONCLUSION

This study demonstrated that functioning assessed through the ICF framework using the PEDI-CAT was associated with clinical and respiratory severity in children and adolescents with cystic fibrosis. Differences were observed across respiratory severity levels, particularly in the Responsibility domain, in which participants with moderate-to-severe impairment presented higher scores.

The PEDI-CAT proved to be a feasible and multidimensional instrument for assessing functioning in this population. Although the 6MWT did not show associations with PEDI-CAT domains in this sample, further longitudinal studies are necessary to better understand the clinical implications of these findings.

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