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ORIGINAL ARTICLE

Adherence to chest physiotherapy, airway clearance techniques and physical exercise by children and adolescents with cystic fibrosis

Larissa Pereira Módolo[a], Luana Bergamim Uliana[a], Fernanda Mayrink Gonçalves Liberato[a], Luana da Silva Baptista Arpini[b], Cíntia Helena Santuzzi[a], Roberta de Cássia Nunes Cruz Melotti[b], Flavia Marini Paro[a]

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[a]Departamento de Educação Integrada em Saúde, Universidade Federal do Espírito Santo (UFES), Vitória, ES, Brazil;

[b]Hospital Estadual Infantil Nossa Senhora da Glória, Vitória, ES, Brazil.

Corresponding author
flamarp@yahoo.com
flavia.paro@ufes.br

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Abstract

Introduction: chest physiotherapy, airway clearance techniques, and physical exercise are associated with better outcomes in cystic fibrosis, but adherence to these recommendations remains a challenge.

Objective: to assess adherence to chest physiotherapy, airway clearance techniques, and physical exercise by children and adolescents with cystic fibrosis and analyze the factors related to adherence.

Methods: retrospective study of children and adolescents with cystic fibrosis from a pediatric referral center in Brazil. Data were collected from electronic records of the multidisciplinary team responsible for caring for these patients at the referral center.

Results: the sample included all 83 patients registered at the referral center, with a mean age of 7.88 ± 4.88 years. Among them, 28.9% did not have weekly chest physiotherapy, 66.3% did not practice physical exercise regularly, and only 38.6% practiced airway clearance techniques from 6 to 7 times a week. Higher adherence to airway clearance techniques was associated with higher mean age ($p<0.01$) and correlated with FEV1 ($r=-0.39$; $p=0.03$), FEF25-75% ($r=-0.36$; $p=0.02$), FEV1/FVC ($r=-0.43$; $p=0.01$) and lower mean Shwachman-Kulczycki Score ($r=-0.34$; $p<0.01$). Higher physical exercise adherence was associated with a higher mean age ($p<0.01$). Individuals using public financing of physiotherapy services had a lower Shwachman-Kulczycki Score ($p=0.02$) than those using non-public services. The metropolitan region had a higher percentage of individuals using non-public services than other regions ($p<0.01$).

Conclusion: adherence to chest physiotherapy, airway clearance techniques and physical exercise was lower than recommendations. Airway clearance techniques and physical exercise adherence increased with age, and airway clearance techniques adherence was associated with the high severity of disease. These results suggest that patients only increase adherence when they get older and the disease worsens, highlighting the need for strategies to increase adherence early.

Keywords: physical therapy, cystic fibrosis, exercise, patient compliance, evidence-based practice.

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Authors summary

Why was this study done?
This research presents new information about adherence to chest physiotherapy, airway clearance techniques and physical exercise among children and adolescents with cystic fibrosis.

What did the researchers do and find?
In our study, we investigated the adherence to chest physiotherapy, airway clearance techniques and physical exercise among children and adolescents with cystic fibrosis. The results showed that the adherence was lower than recommended and increased with age and disease severity.

What do these findings mean?
Considering that chest physiotherapy, airway clearance techniques and physical exercise are recommended for every individual with cystic fibrosis, and poor adherence to them is related to worse outcomes, our findings showed some factors related to poor adherence and highlighted the need for strategies to increase adherence early.

Highlights
The adherence to chest physiotherapy, airway clearance techniques and physical exercise was lower than recommended among the pediatric population with cystic fibrosis.
The adherence increased with age and disease severity.
The results highlighted the need for strategies to increase adherence early.

INTRODUCTION

Cystic fibrosis (CF) is a genetic autosomal recessive disease characterized by variants in the cystic fibrosis transmembrane conductance regulator (CFTR) gene1, which causes epithelial transport abnormalities. The dysfunctions of the CFTR protein are associated with multiorgan impairment and shortened life expectancy.[1] The digestive and the respiratory systems are the most affected, and progressive respiratory impairment is the main cause of morbidity and mortality in CF.[2]

The progression of lung function deterioration in CF is nonlinear and has high variability in the rate of decline over a lifetime, and the rate of decline has slowed over the past several decades because of better treatment.[3]

The increase in life expectancy of individuals with CF has been attributed to several factors, such as greater efficiency in diagnosis,[4-5] evolution of medicines; treatment in specialized centers, the accompaniment of the interdisciplinary team,[2,4,5] and greater knowledge about the importance of treating infections, improving mucociliary clearance in the airways and correcting nutritional deficiencies.[2,4]

Thus, multidisciplinary assistance for individuals with CF is considered fundamental by national5 and international guidelines,[6-8] which include the recommendation of regular and continuous chest physiotherapy, including airway clearance techniques, for all CF patients since diagnosis,[5-8] with daily frequency.[4,5,8]

Chest physiotherapy improves mucociliary clearance,[4,6-9] contributes to preventing infections[4] and postponing the progression of pulmonary disease.[7,8] Furthermore, chest physiotherapy is fundamental to improve cardiovascular fitness and to prevent and treat musculoskeletal complications, among other indications.[7-8] Physical activity and exercise are also recommended[6-8] for every individual with CF, with an exercise frequency of 3-5 times a week and an exercise duration of 20-30 min.[5] However, adherence to these recommendations is an important challenge, and poor adherence is related to worse outcomes.[4] Therefore, studies about the adherence of patients with CF to chest physiotherapy and exercise are important to plan strategies to increase compliance, which has important benefits in the treatment outcomes.

Thus, this study aimed to investigate the adherence to chest physiotherapy, airway clearance techniques, and physical exercise by children and adolescents with cystic fibrosis and to analyze the associations of clinical and demographic variables with adherence.

METHODS

Study Design

This is a cross-sectional retrospective study.

Study Location and Period

Data collection was performed at Hospital Infantil Nossa Senhora da Glória, which is the only pediatric referral center for the treatment of individuals with CF at the state of Espírito Santo (ES), Brazil. Patients registered at the referral center, between May 2018 and March 2019 were included.

Study Population and Eligibility Criteria

The inclusion criteria were children and adolescents, both sexes, up to 18 years old, registered in CF service at the referral center, between May 2018 and March 2019, with a diagnosis of CF, confirmed through sweat test or genetic examination. The exclusion criteria were incomplete records that made data collection impossible. Since this is the only pediatric referral center for CF treatment at the ES state, it is supposed that all patients with a CF diagnosis in the state are registered there.

Data Collection

Data were collected from the medical and multidisciplinary team electronic records in the CF service of the referral center.

In the lung function assessment, we collected the data of the most recent spirometry exam (up to six months), following the procedures according to the Brazilian Society of Pulmonology and Phthisiology criteria.[10] The spirometry test was performed in patients as early as seven years old, capable of completing the test. Therefore, all results regarding lung function include only individuals

over seven years old who can complete the spirometry test.

The following spirometry data were collected: forced expiratory volume in the first second (FEV1), forced vital capacity (FVC), ratio between forced expiratory volume in the first second and forced vital capacity (FEV1/FVC); forced expiratory flow between 25-75% of forced vital capacity (FEF25-75%).

We used the Shwachman-Kulczycki Score (SK) to classify the patients regarding disease severity (< 40 = severe; 41-55 = poor; 56-70 = average; 71-85 = good; and 86-100 = excellent).[11]

The following data were also collected from the physiotherapist's records. This information was part of the standard form prepared by the team of physiotherapists at the service.

1) Is the individual undergoing chest physiotherapy? If the answer was yes, the following data were collected: the weekly frequency, place of chest physiotherapy attendance, and the form of financing access to physiotherapy service (public, health plan/health insurance, or private).

2) Does the patient practice at home the airway clearance techniques that were taught and prescribed by the physiotherapist? If the answer was yes, the weekly frequency and the type of airway clearance techniques were collected. Regarding the children under the age of 6 years old, the parents or guardians were asked whether they had been performing in their child the home airway clearance techniques taught and prescribed by the physiotherapist.

3) Do you practice physical exercise or sports? If the answer was yes, the weekly frequency and the exercise modality were collected.

In the presentation of the results and throughout the article, these three variables were named chest physiotherapy, airway clearance techniques, and physical exercise, respectively.

Data Analysis

The Statistical Package for Social Sciences (IBM SPSS Statistics 20, IBM, Armonk, NY, US) was used for the statistical analysis. Categorical data are presented as absolute and relative frequencies. Continuous variables are shown as mean, standard deviation (SD), minimum, and maximal values. The Shapiro-Wilk test was used to assess data normality. Chi-square and Fisher's exact tests were used for categorical variables. The Mann-Whitney test and Kruskal-Wallis test were used to assess the differences between the groups. To analyze correlations, Spearman's correlation coefficient was used. The correlation strength was determined by the correlation coefficient value (r), according to the following criteria: if $0 < r < 0.3$, there is a weak correlation; if $0.3 < r < 0.6$, there is a moderate correlation; if $0.6 < r < 0.9$, there is a strong correlation; if $0.9 < r < 1$, there is a very strong correlation.12 The significance level adopted was 5%.

Ethical and Legal Aspects of the Research

The Health Sciences Center Research Ethics Committee at the Federal University of ES approved this study (CAAE: 73680617.9.0000.5060).

RESULTS

The sample included all 83 patients registered and treated at the pediatric referral center who were diagnosed with CF. No patients needed to be excluded, considering the previously established criteria.

The mean age of patients was 7.88 ± 4.88 years, ranging from zero years and three months to 17 years and nine months. The average of BMI/I was 16.31 ± 2.07, ranging from 13.20 to 22.50. Most of them were male (54.2%), and 48.2% were white. In the evaluation of lung function, percentages of the predicted values of FEV1, FEF25-75%, and FEF25-75/FVC were below reference values.10 (Table 1). The average SK was 89.82 ± 11.57, ranging from 50 to 100. Therefore, the KS score presented by patients varied from poor to excellent. In the nutritional assessment, there was a higher prevalence of individuals classified as eutrophic (72.3%). Most individuals lived in the Metropolitan Region of ES (60.2%). Details on the clinical and demographic characteristics are in Table 1.

Table 2 shows the results regarding patients' adherence to chest physical therapy, airway clearance techniques, and physical exercise. It was observed that 28.9% of the patients were not seen weekly by physiotherapists, and 41% were seen by physiotherapist only once or twice a week. The region with the highest proportion of children and adolescents with CF treated by a physiotherapist weekly was the metropolitan region. On the other hand, the North Region had the lowest proportion, only 57.1%. Most of the individuals used public financing for chest physiotherapy care (69.5%).

The practice of physical exercise was present in the weekly routine of only 33.7% of the patients. Table 2 presents the weekly frequency of physical exercise.

However, when we consider only the participants from 6 years old (scholar age), 47.2% practiced physical exercise at least once a week.

Regarding airway clearance techniques, most patients (68.7%) reported the practice of them; however, only 38.6% of individuals practiced the techniques with a frequency of 6 to 7 times a week (Table 2).

Furthermore, when only individuals aged from 6 years old or over were included in the analysis (n=53; 63,85%), the percentage of individuals who performed airway clearance techniques increased to 84.9%, but only 50.9% reported practicing them (6-7 times a week).

The most cited modalities of physical exercise were football (28.6%), wrestling (25%), cycling (14.3%), and swimming (14.3%). The airway clearance techniques most frequently cited were high-frequency oral oscillation (91.2%), positive expiratory pressure (43.9%), and breathing patterns (17.5%), without information about what kind of breathing patterns in the records.

Table 3 shows the association between the characteristics of the participants and the variables chest physiotherapy, airway clearance techniques, and physical exercise. There were no characteristics associated with undergoing chest physiotherapy.

Higher adherence to airway clearance techniques was associated with higher mean age ($p < 0.01$), lower average of the lung function variables FEV1 ($p = 0.01$), FEF25-75% ($p = 0.02$), and FEV1 / FVC ($p = 0.01$), and a

lower mean in SK (p <0.01). Regarding physical exercise, greater adherence was only associated with a higher mean age (p < 0.01) (Table 3).

Table 4 presents the correlation coefficients among the studied variables. SK had a moderate negative correlation with age, a moderate positive correlation with FVC, and strong positive correlations with FEV1, FVC, and FEF25-75%. In addition, the weekly frequency of airway clearance techniques had moderate negative correlations with SK score, FEV1, FEV1 / FVC, and FEF25-75%.

Table 5 shows the association between the form of financing physiotherapy care and the other variables. The group of participants with non-public financing (private, health plan and medical insurance) presented higher SK scores (p = 0.02) and a higher percentage of patients residing in the Metropolitan Region (93.3%) than in the group of individuals with public financing (51.3%), p <0.01.

Table 1: Demographic, anthropometric, and clinical characteristics of children and adolescents with cystic fibrosis. Vitoria/Espírito Santo, Brazil, 2018-2019. (N= 83)

Characteristics	
Age(n=83), y and m: mean (SD); [min-max]	7.88 (4.89); [0y3m-17y9m]
From 0y3m to 5y11m	30 (36.15)
From 6y to 11y11m	32 (38.55)
From 12y to 17y9m	21 (25.30)
BMI, kg/m2: mean (SD); [min-max]	16.31 (2.07); [13.20-22.50]
Sex (n=83), Male/ Female, n (%)	45 (54.2)/ 38 (45.8)
Race / Skin color (n=83), n (%)	
White	40 (48.2)
Black	38 (45.8)
Brown	5 (6.0)
Lung Function (% of predicted), (n=46), mean (SD)	
FEV1	79.14 (29.41)
FVC	92.74 (24.64)
FEV1/FVC	81.71 (13.51)
FEF25-75%	64.26 (37.51)
FEF25-75/FVC	63.55 (30.02)
Nutritional status (n=83), n (%)	
Underweight and severe underweight	9 (10.8)
Eutrophic under nutritional risk	12 (14.5)
Eutrophic	60 (72.3)
Overweight	2 (2.4)
Shwachman-Kulczycki Score, mean (SD); [min-max]	89.82 (11.57); [50-100]
Region (n=83), n (%)	
Metropolitan	50 (60.2)
Central	13 (15.7)
South	10 (12.0)
North	7 (8.4)
Municipalities of other states	3 (3.6)

SD, standard deviation; min, minimum value; max, maximum value; y: years; m: months; BMI, body mass index; FEV1, forced expiratory volume in the first second; FVC, forced vital capacity; FEV1/ FVC, FEV1/FVC ratio; FEF25-75%, forced expiratory flow at 25-75% of CVF; FEF25-75/CVF, FEF25-75/FVC ratio.
Source: author

Table 2: Descriptive analysis of patients undergoing chest physiotherapy, physical exercise, and airway clearance techniques (N = 83).

Characteristics	n (%)
Individuals undergoing chest physiotherapy care regularly	59 (71.1)
Frequency of chest physiotherapy care (times a week)	
1 - 2	34 (41)
3 - 5	15 (18.1)
6 - 7	8 (9.6)
Without information	2 (2.4)
Macro-regions of residence in State of ES	
Metropolitan	37 (74.0)
Central	7 (70.0)
South	9 (69.2)
North	4 (57.1)
Municipalities of other states	2 (66.7)
Form of financing access to physiotherapy service	
Public	41 (69.5)
Health plan / health insurance	5 (8.5)
Private	10 (16.9)
Without information	3 (5.1)
Adherence to physical exercise	28 (33.7)
Frequency of physical exercise (times a week)	
1 - 2	13 (15.7)
3 - 5	9 (10.8)
6 - 7	6 (7.2)
Adherence to airway clearance techniques	57 (68.7)
Frequency of airway clearance techniques (times a week)	
1 - 2	3 (3.6)
3 - 5	4 (4.8)
6 - 7	32 (38.6)
Without information	2 (2.4)

ES, Brazilian state of Espírito Santo
Source: author.

Table 3: Association of participants' characteristics and the variables: chest physiotherapy, airway clearance techniques, and physical exercise.

Characteristics	Chest physiotherapy		p	Airway clearance techniques		p	Physical exercise		p
	No	Yes		No	Yes		No	Yes	
Age, mean (SD)	9.33 (5.42)	7.29 (4.58)	0.09[b]	5.04 (4.49)	9.18 (4.54)	<0.01[b]	6.73 (4.99)	10.14 (3.86)	<0.01[b]
Race, n (%) h									
White	9 (37.5)	31 (52.5)	0.21[a]	12 (46.2)	28 (49.1)				
0.80a	23 (41.8)	17 (60.7)	0.10[a]						
Black / Brown	15 (62.5)	28 (47.5)		14 (53.8)	29 (50.9)		32 (58.2)	11 (39.3)	
Lung Function (% predicted), mean (SD)									
FEV1	85.50 (26.15)	75.23 (31.08)	0.27[c]	101.17 (17.13)	75.47 (29.56)	0.01[c]	75.45 (27.80)	82.50 (31.05)	0.37[b]
FVC	95.56 (21.84)	91.00 (26.48)	0.56[c]	107.17 (19.01)	90.33 (24.86)	0.12[c]	89.45 (22.89)	95.73 (26.29)	0.41[c]
FEF25-75%	73.44 (35.28)	58.62 (38.39)	0.21[c]	93.83 (16.86)	59.33 (37.86)	0.02[b]	60.80 (38.48)	67.41 (37.23)	0.64[b]
FEV1/FVC	86.06 (11.02)	79.04 (14.38)	0.08[b]	93.00 (4.69)	79.83 (13.61)	0.01[b]	81.65 (13.43)	81.77 (13.90)	0.99[b]
S-K, mean (SD)	88.33 (10.90)	90.42 (11.86)	0.24[b]	95.58 (5.89)	87.19 (12.57)	<0.01[b]	90.09 (11.41)	89.29 (12.07)	0.95[b]

SD, standard deviation; FEV1, forced expiratory volume in the first second; FVC, forced vital capacity; FEF25-75%, forced expiratory flow at 25-75% of CVF; FEV1/ FVC, FEV1/FVC ratio; S-K, Shwachman-Kulczycki Score; p-value < 0,050; a Chi-square test; b Mann-Whitney test; cT-test for means.
Source: author.

Table 4: Chest physiotherapy, airway clearance techniques, and physical exercise: correlations with patients' characteristics

Characteristics	r	p
Shwachman-Kulczycki Score		
Age	-0.35*	<0.01
Lung Function		
FEV1	0.66*	<0.01
FVC	0.63*	<0.01
FEV1/FVC	0.49*	<0.01
FEF25-75%	0.62*	<0.01
Chest physiotherapy (weekly frequency)		
Shwachman-Kulczycki Score	0.06	0.57
Lung Function		
FEV1	-0.15	0.34
FVC	-0.09	0.55
FEV1/FVC	-0.22	0.17
FEF25-75%	-0.18	0.27
Physical exercise (weekly frequency)		
Shwachman-Kulczycki Score	0.02	0.87
Lung Function		
FEV1	0.28	0.07
FVC	0.28	0.07
FEV1/FVC	0.09	0.58
FEF25-75%	0.20	0.19
Airway clearance techniques (weekly frequency)		
Shwachman-Kulczycki Score	-0.34*	0.01
Lung Function		
FEV1	-0.39*	0.03
FVC	-0.36	0.05
FEV1/FVC	-0.43*	0.02
FEF25-75%	-0.43*	0.02

r, correlation coefficient; FEV1, forced expiratory volume in the first second; FVC, forced vital capacity; FEV1/ FVC, FEV1/FVC ratio; FEF25-75%, forced expiratory flow at 25-75% of CVF; * p-value < 0.050.
Source: author.

Table 5: Association between participants' characteristics and forms of access to physiotherapy.

Characteristics	Forms of financing access to physical therapy service		p
	Public	Non-public	
Age, mean (SD)	7.34 (4.39)	6.93 (5.02)	0.66[a]
Race, n (%)			0.07[b]
White	19 (46.3)	11 (73.3)	
Black / Brown	22 (53.7)	4 (26.7)	
Chest physiotherapy (wk)	2.4 (1.22)	3.93 (2.52)	0.13[a]
Airway clearance techniques (wk)	4.18 (3.35)	3.83 (3.38)	0.90[a]
Physical exercise (wk)	1.24 (2.22)	1.33 (1.84)	0.66[a]
Lung Function (% of pred.), mean (SD)			
FEV1	67.40 (29.56)	98.25 (17.50)	0.06[a]
FVC	84.35 (24.37)	110.50 (20.17)	0.09[a]

Continuation - Table 5: Association between participants' characteristics and forms of access to physiotherapy.

Characteristics	Forms of financing access to physical therapy service		p
FEV1/FVC	76.40 (15.37)	87.75 (5.12)	0.24[a]
FEF25-75%	50.50 (39.29)	83.25 (15.59)	0.11[a]
S-K, mean (SD)	88.17 (13.03)	95.33 (6.94)	0.02[a]
Region, n (%)			
Metropolitan	20 (51.3)	14 (93.3)	<0.01[c]
Other regions	19 (48.7)	1 (6.7)	

SD, standard deviation; wk, times a week; FEV1, forced expiratory volume in the first second; FVC, forced vital capacity; FEV1/ FVC, FEV1/FVC ratio; FEF25-75%, forced expiratory flow at 25-75% of CVF; S-K, Shwachman-Kulczycki Score; p-value < 0.05; a Mann-Whitney test; b Chi-square test; c Fisher's exact test.
Source: author.

DISCUSSION

The results showed low adherence by children and adolescents with CF to chest physiotherapy, physical exercise, and airway clearance techniques. In fact, 28.9% of participants did not have chest physiotherapy care, only 3.6% underwent physiotherapy daily, 38.6% reported daily adherence to airway clearance techniques, and 18% reported adherence to physical exercise three or more times a week.

The low adherence to daily chest physiotherapy is worrisome since poor compliance is correlated with worse outcomes, with accelerated decline in lung function and the need for increased interventions with antibiotics.[4] A systematic review concluded that chest physiotherapy has a short-term effect on the increase in secretion transport and elimination in patients with CF,[9] corroborating the recommendation that physiotherapy should be done daily[7]. So, increasing physiotherapy adherence and access is a cornerstone to improve the outcomes of patients with CF.

However, many individuals with CF and their families face challenges with the routine of journeys to specialized treatment centers, sometimes placed tens or even hundreds of kilometers away from their residences,[13] which can seriously compromise adherence. In the present study, the North region, which is the most distant region from the referral center, had the lowest proportion of individuals with CF who reported undergoing chest physiotherapy, only 57.1%, while the Metropolitan region had the highest adherence (74%). In addition, although 41.2% of patients from the Metropolitan region used non-public physiotherapy services, 99.5% of patients from other regions reported using public physiotherapy services, which may be related to socioeconomic differences and access to health services between regions. There is a need to study adherence to CF therapies and formulate strategies to increase compliance and overcome the barriers.[14, 15] Undoubtedly, public physiotherapy services for children and adolescents with CF in their municipality of residence is a low cost, simple, and fundamental strategy to overcome barriers and increase adherence.

Telephysiotherapy, which can include telemonitoring, telecoaching, and telerehabilitation, has been growing used in physiotherapy[16], and technological tools have been increasingly applied to improve health education[17]. So, it could be an alternative to increasing physiotherapy access for some patients with CF living in more distant regions.

In a study with the CF pediatric population aged from 6 to 17 years old, 59% of patients reported performing airway clearance techniques daily.[18] Our sample showed lower adherence, as only 38.6% of patients reported performing airway clearance techniques 6 to 7 days a week. However, our sample also included individuals aged under 6 years, which can explain the lower adherence, as our results showed a higher average age among patients who reported practicing airway clearance techniques (9.18 ± 4.54 years old) versus patients who did not practice them (5.04 ± 4.49 years old), p<0.01.

It is important to assess adherence in CF children younger than 6 years old. Adherence at this age depends basically on parents or guardians, who should ensure that the child performs the techniques prescribed by physiotherapists. It is worth mentioning that airway clearance techniques with daily frequency are recommended since CF diagnosis.[2,4,5,7] Thus, educational strategies to increase awareness about the disease and parent training could be tools to increase adherence in this age.[14]

Adults with CF have shown higher adherence rates than the pediatric population,[19, 20] which also suggests that adherence increases with age. Only one contradiction was found about the positive association between adherence and age. A study conducted in Spain found that adherence to treatment worsened with age and the severity of the disease; however, the same study mentioned that adherence increased with the perception of the importance of treatment and its impact on quality of life.[20] It is important to note that this kind of perception and awareness has a trend to increase with the increase in age and education about the disease.

SK is widely used for monitoring the severity of CF, adequately reflecting lung function impairment.[11] In the present study, SK presented a negative correlation with age and a positive correlation with lung function, which corroborates well-established data from the literature. [21, 22]

Moreover, the frequency of airway clearance techniques had a negative correlation with SK and with variables of lung function, which means greater adherence

among individuals presenting more severe illness. Feiten *et al*.[18] and Arias *et al*.[21] reported no significant results when relating adherence to SK. In contrast, SK and pulmonary function were significantly lower in patients with high adherence in other studies.[19,20] The hypothesis for this association is that the severity of illness worsens respiratory symptoms, increasing the perception of airway clearance techniques benefits. Corroborating this hypothesis, the body mass index was negatively correlated with airway clearance techniques adherence in adults with CF in another study.[23] Low body weight in CF has been related to worse lung function[24] and quality of life.[25] In addition, low body weight increases the impact of respiratory symptoms on the quality of life,[26] which probably makes patient aware of their need for airway clearance techniques, increasing adherence.

It is a concern that patients with less severe disease had poor adherence to airway clearance techniques because they likely will increase adherence only when irreversible lung damage has occurred.[14,19] Therefore, health education has a key role in awareness of patients with less severe disease of the importance of adherence to improve their prognosis. Some strategies suggested to increase adherence to treatment were telephone monitoring, home visits,[27] education, self-monitoring, reminders, parent training, motivational interviewing, and behavioral therapy.[14] In children with CF, telephone monitoring and home visits were demonstrated to improve treatment adherence, lung function, and quality of life.[28]

Increasing early diagnosis is another important strategy to ensure early specialized attention and adherence to physiotherapy from the beginning of life, improving the prognosis of the disease. Primary Health Care plays a fundamental role in this process. The CF newborn screening algorithm used in Brazil is available in the public health system freely for all newborns, together with screening for other diseases. If screening is positive, diagnosis tests are performed to confirm or rule out CF. Immediately after diagnosis, CF requires early multidisciplinary management.[5] However, studies in different Brazilian states, including Espírito Santo, have shown that a relevant proportion of patients are diagnosed later[28, 29]. So, strategies for health promotion and education are necessary.

The techniques most recommended to CF patients by physiotherapists from 9 referral centers in Brazil, including this referral center, were huffing (61.1%), high-frequency oral oscillation (52.0%) and positive expiratory pressure (45.3%).[30] Similarly, in the present study, the most cited techniques by patients were high-frequency oral oscillation (91.2%), positive expiratory pressure (43.9%), and breathing patterns (17.5%). Adherence can vary depending on the airway clearance technique used,[14] and 'do not enjoy technique' was reported as one of the reasons for low adherence.[18]

Guidelines for the Diagnosis and Treatment of CF recommend the practice of aerobic and anaerobic exercises, with a frequency of 3 to 5 times a week.[5] Despite this, in our sample, only 33.7% of patients performed physical exercise weekly, and only 18% performed physical exercise three or more times a week. Factors such as perception of the importance of treatment, immediate effects, self-esteem, and social and family support influence adherence to habitual physical activity.[31] In the present study, it was also observed that physical exercise adherence increased with age, possibly because the study included children under six years old, in preschool age, therefore still without access to exercises and sports activities in schools. Considering only individuals aged six years old, the adherence increased to 47.2%, which was still too low.

In individuals with CF, the beneficial effect of physical training on exercise capacity, lung function, and quality of life was reported in a systematic review.[32] In addition, a previous study reported an association between cardiopulmonary exercise testing variables at the ventilatory threshold and CF exacerbations, concluding that percentage $\dot{V}O_2$ at the ventilatory threshold could serve as a complementary variable to monitor exacerbations in people with CF.[33] In addition, the reduction in exercise capacity is associated with a decline in pulmonary function and life expectancy.[6]

Patients who used public physical therapy services had lower SK values than individuals using non-public services. This result suggests greater severity of patients using public physiotherapy services than nonpublic patients. This may be related to the social status of patients. Low socioeconomic status has a negative effect on health in CF,[34, 35] and it has been reported that people with CF from socioeconomically disadvantaged backgrounds die younger than those in more advantaged situation.[35] This issue deserves other studies but reinforces the need to offer public physiotherapy services to all patients with CF in their municipalities. It has been recommended to healthcare teams implement interventions to reduce disparities at CF centers and study their impact.[34]

The retrospective design of this study might present some limitations, like imprecision of some information and bias of selection and memory. However, this risk of bias was reduced because the cystic fibrosis program team of this hospital is used to conduct many research projects and the charts are very precise and complete in their information, and these patients were routinely assessed at intervals ranging from 15 days to 3 months. Although the sample can be considered relatively small and restricted to a single center, it is representative of the entire population of a Brazilian State, which has 3,833,486 inhabitants, because the study included the entire population of children and adolescents with CF registered in the only state pediatric referral center of the State, where all patients with CF in the state are registered.

CONCLUSION

In conclusion, the study showed adherence to chest physiotherapy, airway clearance techniques, and physical exercises too lower than recommended. Physical exercise and airway clearance techniques adherence increased with age, and higher respiratory exercise adherence was associated with higher severity of disease. These results suggest that the patients only increase adherence when they get older and the disease worsens, highlighting the need for strategies to increase early adherence.

Author Contributions

All authors contributed to the manuscript. Literature search: LPM, LBU, FMGL, LSBA, RCNCM and FMP

Data collection: LPM, LBU, FMGL, LSBA, RCNCM and FMP Study design: LPM, LBU, FMGL, LSBA, RCNCM and FMP Analysis of data: LPM, LBU, FMGL, LSBA, CHAS and FMP Manuscript preparation: LPM, LBU and FMP Review of manuscript: LPM, LBU, FMGL, LSBA, RCNCM, CHAS and FMP

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Conflicts of Interest

The authors report no conflict of interest.

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Resumo

Introdução: fisioterapia respiratória, técnicas de desobstrução das vias aéreas e exercícios físicos estão associados a melhores prognósticos na fibrose cística (FC), contudo a adesão dos pacientes é um desafio.

Objetivo: avaliar a adesão das crianças e adolescentes com FC à fisioterapia respiratória, às técnicas de desobstrução das vias aéreas e ao exercício físico e analisar fatores relacionados à adesão.

Método: estudo retrospectivo que incluiu crianças e adolescentes com FC de um centro de referência pediátrico brasileiro. Os dados foram coletados dos registros eletrônicos da equipe multidisciplinar responsável pelo atendimento desses pacientes no centro de referência.

Resultados: a amostra incluiu os 83 pacientes cadastrados no centro de referência, com média de idade de 7,88 ± 4,88 anos, entre os quais, 28,9% não faziam fisioterapia respiratória semanalmente, 66,3% não praticavam exercícios físicos regularmente, e apenas 38,6% praticavam as técnicas de desobstrução das vias aéreas de 6 a 7 vezes por semana. Adesão às técnicas de desobstrução das vias aéreas apresentou associação positiva com idade ($p<0,01$) e correlação negativa com: VEF1 ($r=-0,39$; $p=0,03$), FEF25-75% ($r=-0,36$; $p=0,02$), VEF1/CVF ($r=-0,43$; $p=0,01$) e escore de Shwachman-Kulczycki ($r=-0,34$; $p<0,01$). Adesão ao exercício físico associou-se positivamente à idade ($p<0,01$). A utilização de financiamento público de serviços de fisioterapia associou-se a menor escore de Shwachman-Kulczycki ($p=0,02$). A região metropolitana apresentou o maior percentual de indivíduos que utilizam serviços de fisioterapia não públicos ($p<0,01$).

Conclusão: a adesão à fisioterapia respiratória, às técnicas de desobstrução das vias aéreas e ao exercício físico foi inferior às recomendações. A adesão às técnicas de desobstrução das vias aéreas e ao exercício físico aumentou com a idade, e a adesão às técnicas de desobstrução também se associou à gravidade da doença, o que reforça a necessidade de estratégias para aumentar a adesão precocemente.

Palavras-chave: fisioterapia, fibrose cística, exercício, cooperação do paciente, prática clínica baseada em evidências.