

ORIGINAL RESEARCH

Respiratory physical therapy techniques recommended for patients with cystic fibrosis treated in specialized centers



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KEYWORDS

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Abstract

Background: Respiratory therapy is a part of the treatment of patients with cystic fibrosis (CF). However, there is no information about the main factors influencing the recommendation of the use of different techniques or devices by physical therapists from different specialized centers.

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Physical therapy modalities;
Pulmonary function

Objective: To determine the respiratory therapy techniques recommended for the treatment of patients with CF seen in specialized treatment centers in Brazil.

Methods: This is a descriptive study including a sample of Brazilian CF specialized treatment centers. Data on demographics, spirometric variables, and recommended respiratory therapy treatment techniques were collected.

Results: Twelve specialized treatment centers with a total of 974 patients were included (13.5 ± 11.3 years old and FEV_1 (%) 75.7 ± 27.9). The most recommended techniques were huffing (61.1% of patients), high frequency oral oscillation (HFOO) (52.0%), and positive expiratory pressure (PEP) (45.3%). Most often, recommendation was to perform treatment once (54.8% of patients) or twice (34%) a day. There was great variability in the recommendation among the different states. When data were separated by age, there was a predominance of performing conventional and manual techniques in infants and preschool children. There were no significant variations according to pulmonary function. Based on the literature, techniques based on volume, huffing, and PEP were most prevalently performed in international centers.

Conclusion: The most recommended treatment techniques for patients with CF in Brazil are huffing, HFOO, and PEP, followed by volume-based techniques. There were geographical variations in the preferred treatment techniques, as well as based on patient age, but not based on the level of pulmonary function.

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Introduction

Cystic fibrosis (CF) is a genetic disease that affects multiple systems leading to changes in the exocrine glands and causing the production and retention of thick secretions.¹ In the respiratory tract, these secretions induce recurrent infections and inflammation, bronchiectasis, increased airway resistance, pulmonary hyperinflation, and, as a consequence, progressive decline in lung function.² Although it is a multisystemic disease, pulmonary complications are primarily responsible for high rates of morbidity and mortality.³

According to data from the Brazilian Registry of CF, the prevalence of CF in Brazil varies according to ethnicity and region of birth, being less prevalent in Afro-descendants and affecting 1/2500 live births in the Caucasian population.⁴ Patients diagnosed early and receiving lifelong treatment have shown an increase in longevity with those born after the 1990s, having an average life expectancy greater than 40 years.⁵

Treatment should start as early as possible, be individualized, and consider factors such as severity and systems affected.^{6,7} Respiratory therapy has historically been a part of the management of patients with CF and evidence in the literature supports its recommendation and benefits.⁸ Respiratory physical therapy devices and techniques provide an increase in the transport of mucus in the airways, contributing to an improvement in mucociliary clearance.⁹ Thus, there is a consensus that airway clearance is essential for patients with CF to slow-down damage and progression of lung disease.¹⁰

In the past, few resources were available for airway clearance and conventional respiratory physical therapy

was basically composed of chest percussion, vibration, and postural drainage.¹¹ Currently, devices and techniques for the removal of bronchial secretions include: expiratory flow acceleration (EFA), thoracic compression, autogenic drainage (AD), active cycle of breathing (ACB), and positive expiratory pressure systems, either using orifice (PEP), spring-load (EPAP), or water seal resistors.¹² Evidence shows that these devices and techniques are all equally effective for patients with CF, although there is an increasing tendency for patients to prefer self-administered treatment approaches, facilitating independence.

Although these devices and techniques are recognized in the treatment of patients with CF, there is still no consensus on which to use in clinical practice, with apparent heterogeneity of practice in different specialized treatment centers in Brazil. Countries such as the United Kingdom, Canada, Ireland, New Zealand, and the United States include in their annual registry of patients with CF the devices and techniques of respiratory therapy most used by patients, which allows to determine the adequacy of treatment for each patient.¹³⁻¹⁷ Therefore, a nationwide survey would contribute to increase knowledge on physical therapy management and possibly help establish a consensus for the management of patients with CF in Brazil.

The objective of this study was to determine the devices and techniques of respiratory physical therapy used in patients with CF in Brazilian specialized treatment centers. In addition, we sought to determine if therapy choices were influenced by age and severity of pulmonary dysfunction. Finally, we sought to compare practice in Brazil with those in other countries.

Methods

An observational descriptive study was conducted to determine the respiratory physical therapy devices and techniques prescribed for patients with CF seen at specialized treatment centers in Brazil. All registered CF centers recognized by the Brazilian Cystic Fibrosis Group were considered potential participating centers ($n = 21$). Contact was performed by electronic mail and/or telephone on at least 2 occasions. Failure to respond or incomplete transmission of data were considered an exclusion criterion.

After a center accepted to participate in the study, the physical therapists responsible for patient care were asked to complete a table in an Excel spreadsheet with the following variables of interest for each patient: age, sex, weight, height, forced expiratory volume in the first second in absolute values (FEV_1 absolute), forced expiratory volume in the first second in percentage values ($FEV_1\%$), respiratory physical therapy devices and techniques prescribed, and number of times a day and number of times a week patients were recommended to use these treatments.

The following categories were used to analyze data: conventional respiratory physical therapy techniques (postural drainage, tapping/percussion, vibration/vibrocompression); manual techniques (passive expiratory chest compression (TEMP), EFA, retrograde rhinopharyngeal clearance (RRC), retrograde rhinopharyngeal clearance with saline instillation (RRCI), thoracic self-compression, chest compression, slow and prolonged expiration, re-expansive maneuvers); volume-based techniques (autogenic drainage (AD), modified AD, assisted AD, forced expiratory technique (FET), ACB; technique of huffing; positive expiratory pressure devices – EPAP, PEP, water seal; high frequency oral oscillators (HFOO) (Flutter®, Shaker®, Acapella®); noninvasive ventilation (NIV); ventilatory exercises with or without breathing devices (diaphragmatic breathing training, prolonged expiratory breathing exercises, forced exhalation exercises, flow incentive spirometry – respiron®, pulmonary expansion exercises, diaphragmatic breathing, blowing exercises); other techniques (thoracic manipulation, abdominal support, mechanical chest vibration, intermittent positive pressure breathing (IPPB), airway suction); no procedures. The use of coughing was not categorized, as it is part of routine physical therapy care to recommend its performance during treatment, independent of the devices and techniques used.

Pulmonary function was determined based on $FEV_1\%$ and categorized as normal ($FEV_1 > 80\%$), mild ($FEV_1 = 79\text{--}60\%$), moderate ($FEV_1 = 59\text{--}40\%$), and severe ($FEV_1 \leq 39\%$), following the American Thoracic Society (ATS) guideline. Patients were also categorized according to the Brazilian Society of Pediatrics as infants (0–2 years), preschoolers (2–4 years), schoolchildren (5–10 years), adolescents (11–19 years), and adults (>19 years). In addition, information from international registries and articles^{13–17} specifying the respiratory physical therapy devices and techniques most frequently used for patients with CF was collected.

The study was approved by the University Research Ethics Committee (Pontifícia Universidade Católica do Rio Grande do Sul – PUCRS) under number 49172815.0.0000.5336. The return of the completed electronic file indicated the accep-

Table 1 Characterization of the sample included in the study.

	RS (n = 274)	SC (n = 66)	PR (n = 13)	SP (n = 104)	RJ (n = 98)	MG (n = 147)	ES (n = 136)	GO (n = 31)	PA (n = 102)	Total (n = 974)
Age, years	16.0 ± 9.6	6.8 ± 4.5	29.4 ± 19.1	12.3 ± 7.8	9.8 ± 5.3	6.5 ± 4.6	18.0 ± 17.3	10.3 ± 7.2	18.4 ± 12.8	13.5 ± 11.3
Sex, M (%)	50.7	57.6	23.1	51.9	45.9	57.8	49.3	32.3	58.1	51.5
Height, cm	145.8 ± 29.6	116.4 ± 31.8	149.3 ± 23.9	137.0 ± 28.2	132.7 ± 29.2	113.0 ± 29.5	139.1 ± 30.0	129.5 ± 29.6	149.3 ± 19.1	36.9 ± 19.9
Weight, kg	44.9 ± 19.7	23.4 ± 13.2	57.4 ± 20.4	35.8 ± 18.4	33.5 ± 17.7	22.3 ± 12.3	38.9 ± 21.3	30.8 ± 15.5	46.9 ± 17.3	135.5 ± 31.1
BMI, kg/m ²	19.5 ± 3.5	15.9 ± 2.56	24.7 ± 5.3	17.6 ± 3.8	17.4 ± 3.6	16.1 ± 2.2	18.2 ± 4.0	16.8 ± 3.1	20.1 ± 4.2	18.2 ± 3.8
FEV_1 abs, L	2.11 ± 1.01	1.54 ± 0.68	1.27 ± 0.65	1.82 ± 0.88	1.90 ± 0.76	1.60 ± 0.6	1.92 ± 0.95	1.79 ± 0.91	2.36 ± 1.07	1.9 ± 0.95
$FEV_1\%$	76.2 ± 31.0	75.3 ± 23.2	50.0 ± 28.9	62.9 ± 23.4	77.6 ± 23.0	79.1 ± 24.4	69.3 ± 26.4	73.3 ± 20.4	86.6 ± 27.6	75.7 ± 27.9

Data expressed as mean ± standard deviation. M, male; BMI, body mass index; FEV_1 abs, absolute values of forced expiratory volume in the first second; RS, Rio Grande do Sul; SC, Santa Catarina; PR, Paraná; SP, São Paulo; RJ, Rio de Janeiro; MG, Minas Gerais; ES, Espírito Santo; GO, Goiás; PA, Pará.

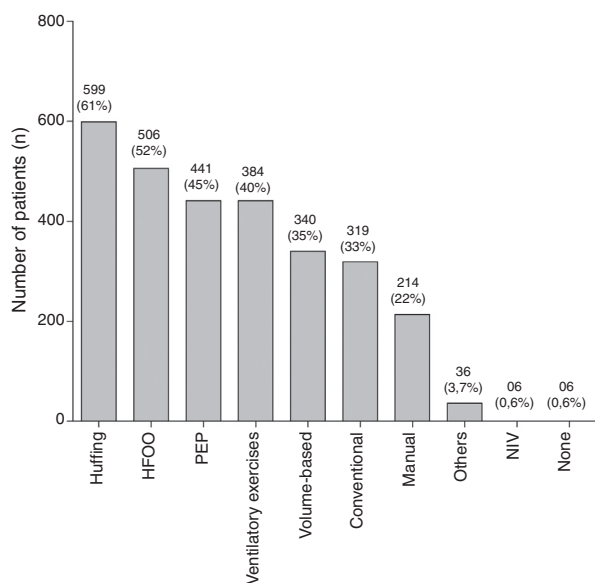


Figure 1 Devices and techniques used for respiratory physical therapy (n = 974). PEP, positive expiratory pressure; HFOO, high frequency oral oscillation; NIV, noninvasive ventilation.

tance of the center to participate in the study. All authors of the present study declared confidentiality regarding the data collected.

For each variable, normality of data distribution was evaluated using the Kolmogorov-Smirnov test. Continuous variables are presented with means and standard deviations while categorical variables are presented as percentages. All analyzes and data processing were performed using SPSS version 18.0 (SPSS Inc., USA).

Results

According to the website of the Brazilian Cystic Fibrosis Study Group, there are 37 registered CF centers. We were about to contact and invite 21 centers, from which 12 sent the required data. Nine centers were excluded due to lack of response or incomplete data transmission. Data from the 12 participating centers included 974 patients, representing 9 states of the country: Rio Grande do Sul (RS), Santa Catarina (SC), Paraná (PR), São Paulo (SP), Rio de Janeiro (RJ), Espírito Santo (ES), Minas Gerais (MG), Goiás (GO), and Pará (PA). These states characterize four regions of Brazil (South, Southeast, Midwest, and North).

Patients had a mean age of 13.5 ± 11.3 years, a mean BMI of $18.2 \pm 3.8 \text{ kg/m}^2$ and 51.5% were male (Table 1). The mean absolute FEV₁ was 1.9 ± 0.95 liters and the mean percentage was $75.7 \pm 27.9\%$, which represents a mild pulmonary impairment.

Huffing (61.1% of the patients), HFOO devices (52.0%), and positive expiratory pressure devices (45.3%) were the most commonly used treatment approaches used in Brazil (Fig. 1). Respiratory therapy was recommended to be performed once a day for 54.8% of patients, twice a day for another 34%, and three or four times per day for 10.4% of patients. Nearly all patients (96.8%) were recommended to perform their therapy daily.

Based on the data for each state, the huffing technique was the most often used approach in RS (83.2%), SC (66.7%), PR (84.6%), SP (68.3%), and PA (98.1%). The HFOO devices were most often used in the states of SC (63.6%), PR (92.3%), SP (63.5%), ES (80.9%), and PA (93.3%). The use of PEP devices was more prevalent in RS (74.5%), RJ (78.6%), and GO (71.0%). Ventilatory exercises, with or without a device, were predominant in SP (85.6%), ES (70.6%), and PA (84.8%). The volume-based techniques had a higher percentage of use in the states of RJ (71.4%), GO (71.0%), and PA (84.8%), and the use of manual techniques was higher in GO (58.1%), finally, conventional techniques were more often used in MG (74.8%) and PA (97.1%) (Table 2).

In the analysis of the use of devices and techniques according to age group, there was predominance of conventional and manual techniques in infants (58.6 and 80.0%, respectively) and preschool children (40.0 and 43.3%, respectively). Volume-based techniques were more often used in adolescents (44.2%) and adults (43.1%). Conversely, the PEP devices were more prescribed for schoolchildren (45.7%) and adolescents (65.2%). Regarding HFOO devices, huffing, and ventilatory exercises, they were more frequently used in schoolchildren (62.2, 49.8, and 45.7%), adolescents (60.2, 42.0, and 65.2%), and adults (56.3, 44.2, and 37.6%), respectively (Table 3).

When data were categorized according to the severity level of pulmonary impairment, no relevant variations were demonstrated (Table 3). In both patients with normal and mild lung function, the most frequent respiratory physical therapy devices and techniques were huffing (73.1 and 72.0%), HFOO (59.1 and 61.9%), and PEP (58.3 and 54.2%), respectively. In patients with moderate pulmonary impairment, the most used techniques were also huffing and HFOO (70.2 and 63.2%), followed by PEP and volume-based techniques (both with 54.3%). In patients with severe pulmonary involvement, there was a predominance of huffing (85.0%), HFOO (57.9%), and volume-based techniques (53.3%).

Table 4 shows the devices and techniques most used in different countries. The volume-based techniques and/or huffing were the most prevalent in Brazil (75.8%), as well as in Ireland (47.5%) and the United Kingdom (28.0%). On the other hand, the PEP devices are more used in Canada (43.7%) and New Zealand (83.3%). In the United States, the most prevalent devices and techniques are in the category of other resources, corresponding to 74.8%, probably because this category includes the high frequency chest wall oscillation (HFCWO).

Discussion

The results showed that most physical therapists of the included Brazilian CF centers recommend the use of devices and techniques such as huffing, HFOO, and PEP. In addition, there is a recommendation for most patients to undergo respiratory therapy seven times a week, once a day.

It is already known that the use of respiratory physical therapy devices and techniques in patients with CF has positive effects.⁹ But, in the last systematic review that analyzed the use of different devices and techniques of respiratory physical therapy in patients with CF, no superiority of any therapeutic approach was identified.¹¹ Despite

Table 2 Recommended physical therapy devices and techniques for each state.

	RS (n=274)	SC (n=66)	PR (n=13)	SP (n=104)	RJ (n=98)	MG (n=147)	ES (n=136)	GO (n=31)	PA (n=98)
Conventional	9.5	10.6	69.2	0	26.5	74.8	24.3	19.4	97.1
Manual	10.2	40.9	0	39.4	1.0	40.1	0.7	58.1	37.1
Volume-based	24.5	30.3	0	19.2	71.4	35.4	0	71.0	84.8
PEP	74.5	47.0	7.7	1.0	78.6	18.4	30.1	71.0	35.2
HFOO	16.4	63.6	92.3	63.5	49	57.1	80.9	3.2	93.3
Ventilatory exercises	0	47.0	0	85.6	1.0	49.0	70.6	22.6	84.8
Huffing	83.2	66.7	84.6	68.3	2.0	21.1	69.9	45.2	98.1
NIV	0.4	0	0	0	0	3.4	0	0	0
Others	0	7.6	0	27.9	0	0.7	0.7	0	0
None	2.2	0	0	0	0	0	0	0	0

Data expressed as a percentage of patients using each technique. The total for each column exceeds 100% as more than one technique is typically recommended. PEP, positive expiratory pressure; HFOO, high frequency oral oscillation; NIV, noninvasive ventilation; RS, Rio Grande do Sul; SC, Santa Catarina; PR, Paraná; SP, São Paulo; RJ, Rio de Janeiro; MG, Minas Gerais; ES, Espírito Santo; GO, Goiás; PA, Pará.

Table 3 Respiratory physical therapy devices and techniques recommended according to the patient's age group and severity level of pulmonary dysfunction.

	Conventional	Manual	Volume-based	PEP	HFOO	Huffing	Ventilatory exercises	NIV	Others	None
Age (years)										
Infants (0–2) - n=70	58.6	80.0	8.6	8.6	11.4	11.4	4.3	0	10.0	0
Preschoolers (2–4) - n=120	40.0	43.3	12.5	25.8	24.2	23.3	22.5	0	12.5	0
Schoolchildren (5–10) - n=267	39.3	13.9	34.8	45.7	62.2	49.8	61.8	0.7	3.4	0
Adolescents (11–19) - n=319	21.3	14.1	44.2	65.2	60.2	42.0	68.3	1.3	1.3	1.3
Adults (>19 years) - n=197	28.4	11.7	43.1	37.6	56.3	44.2	91.4	0	0.5	1.0
Pulmonary Function (FEV ₁ %) ^a										
Normal (>80) - n=375	28.9	10.6	37.8	58.3	59.1	73.1	41.7	0	0.6	1.4
Mild (79–60) - n=118	19.5	9.3	44.1	54.2	61.9	72.0	48.3	1.7	0	0
Moderate (59–40) - n=94	22.3	18.1	54.3	54.3	63.8	70.2	45.7	0	1.1	0
Severe (<39) - n=107	29.9	18.7	53.3	51.4	57.9	85.0	42.1	3.1	2.8	0

Data expressed as a percentage of patients using each technique. The total for each row exceeds 100% as more than one technique is typically recommended. PEP, positive expiratory pressure; HFOO, high frequency oral oscillation; NIV, noninvasive ventilation.

^a Pulmonary function is based on the percentage of forced expiratory volume in the first second (FEV₁).

Table 4 Respiratory physical therapy devices and techniques used in different countries.

	Brazil (n=974)	UK ^a (n=6372)	Canada (n=3946)	Ireland (n=1133)	USA (n=29497)	NZ (n=358)
Conventional	32.8	4.0	28.4	-	7.9	≅ 44.6
Manual	22.0	-	-	-	-	-
Volume-based/huffing	75.8	28.0	≅ 8.0	≅ 47.5	0.8	≅ 22.3
PEP	45.3	16.0	43.7	39.9	8.8 ^c	≅ 83.3
HFOO	52.0	23.0	7.4	≅ 29.3	8.8 ^c	≅ 20.8
Ventilatory exercises	38.5	-	-	-	-	-
NIV	0.6	-	-	5.4	2.8	-
Others	3.7	2.0	≅ 14.0	1.8 ^b	74.8 ^b	≅ 2.4 ^b
None	0.6	10.0	7.4	-	2.8	≅ 17.2

Data expressed as a percentage of patients using each technique. UK, United Kingdom; USA, United States of America; NZ, New Zealand. PEP, positive expiratory pressure; HFOO, high frequency oral oscillation; NIV, noninvasive ventilation.

^a Exercise was reported for 16% of patients and 2% was considered as missing data.

^b Represents the use of high frequency chest-wall oscillation (The Vest[®]).

^c The use of PEP and HFOO was considered in the same category.

the results of that review, there is evidence to suggest that the use of PEP may be more effective than conventional techniques, slowing-down the long-term decline in lung function,¹⁸ and reducing pulmonary exacerbations compared to HFCWO use.¹⁹ Another study suggests that the beneficial effects of HFOO techniques are more apparent in the short term compared to PEP²⁰ and, the use of Flutter may improve lung function of patients who are hospitalized.²¹ Furthermore, evidence demonstrates that huffing may favor expectoration, with increased sputum weight and improved airway clearance in patients with CF.^{22,23} Because some therapies have very specific effects, physical therapy resources and techniques can be optimized when used in association.^{24,25}

According to the Brazilian guidelines for diagnosis and treatment of patients with CF, it is indicated that respiratory physical therapy should be performed once a day, every day.¹² The results presented here have confirmed that physical therapists are prescribing airway clearance as recommended. However, when in the presence of exacerbation, the physical therapist must adjust the number of sessions required according to the patient's clinical condition.²⁶

When analyzing devices and techniques most used in the different states of the country, it was possible to identify a great amount of heterogeneity. In the states of MG (74.8%) and PA (97.1%), for example, there is predominance of conventional techniques, whereas in SP these techniques were not recommended. In the same way, ventilatory exercises were little used in RJ and were not recommended to any patient in the states of PR and RS, while in SP, this technique was recommended for 85.5% of the patients. Also in SP, 27.9% of the patients were instructed to use devices and techniques classified in the category "other resources," which did not occur in other states. To date, there is no evidence available in the literature that demonstrates regional differences in the recommendations or use of respiratory physical therapy devices and techniques, which hinders discussion and a better understanding of the significance of these findings. It is possible to speculate that the variability found might be associated with cultural and training differences of physical therapists. Also, important socioeconomic factors may limit prescription of certain devices and techniques in some regions.

When recommendations were analyzed according to the age group, a greater predominance on the use of conventional and manual techniques was identified for infants and preschoolers. This may be justified by the difficulty of using techniques that provide more autonomy at this age,²⁷ leading to the selection of techniques aimed at increasing the expiratory flow and clearing the airways, in accordance with the peculiarities of the anatomy and physiology of babies and children.²⁷ In contrast, in schoolchildren, adolescents, and adults, the most indicated devices and techniques were HFOO, huffing, and ventilatory exercises, respectively. It is important to emphasize that these therapies are self-administered, promoting greater independence and practicality, which promotes adherence to treatment. Thus, in part, these strategies aim for higher rates of adherence to treatment.²⁸ Considering that CF is a progressive disease, the older the patients, the greater the expected progression of lung disease.²⁹ Thus, it is possible that the age group may have a direct relation with the choice of the

physical therapeutic intervention, aiming to recommend the therapy that best matches the patient's needs and practical use.

The level of severity of pulmonary function did not affect the choice of respiratory physical therapy devices and techniques with huffing, HFOO, and PEP, most often used, independent of pulmonary function. In addition, it is possible that the mild impairment of pulmonary function is responsible for the small percentage of non-invasive ventilation (NIV) recommendation, since this resource is used as adjuvant in the performance of respiratory physical therapy in patients with severe pulmonary impairment. A study in the UK has shown that individuals with mild lung disease have a tendency to use only physical exercise as part of their routine therapy, while patients with severe lung disease are the most likely to use NIV.¹³

When comparing the devices and techniques of respiratory physical therapy most recommended in Brazil with international registries, those using volume-based and/or huffing and PEP devices were the most common in the registries. New Zealand (NZ) and Ireland documents address the techniques used by children/adolescents separately from adults, with data demonstrating that PEP is the most commonly used device for both children/adolescents and adults in NZ.¹⁵ However, in Ireland, most children/adolescents use HFOO, while adults use volume-based techniques.¹⁶ Brazil was the only country that presented a category of ventilatory exercises, however, most international registries present a category of "other resources," in which ventilatory exercises may be present. Regarding NIV, the indication was reported only in Brazil and Ireland, with a small percentage of patients. In the United States,¹⁷ most patients use other resources, such as the HFCWO (*The Vest*®), which is not very common in Brazil, but appears as a secondary option in other registries.^{13,15,16} The low rates of use of this device in Brazil may be associated with its high cost. In addition, studies have already suggested that the use of HFCWO does not result in additional benefits when used as a primary respiratory physical therapy intervention in comparison to other techniques.³⁰ Physical exercise is also addressed in the literature as an intervention that assists in airway clearance. In the New Zealand registry, for example, 35.5% of adult patients used exercise as an adjuvant for clearing the airways.¹⁵ In the present study, physical exercise was not considered as a device or technique for respiratory physical therapy, but it is known that its indication is routine as an adjunct to treatment, without replacing the indication for specific respiratory physical therapy.

The present study is not without limitations. No data were obtained from 9 of the 21 treatment centers of interest, with no data from the northeastern region of the country, which may not represent a large data set as the disease is more prevalent in the southern and southeastern regions. Second, it is possible that the low social or economic status of patients would prevent the physical therapist to recommend a specific device/technique, which would potentially influence some of the results. Finally, the comparison with devices/techniques used in different countries may have been influenced by the different categories and nomenclatures used, as well as factors such as accessibility and cultural differences.

Conclusion

The results indicate that the most recommended respiratory physical therapy techniques for patients with CF in Brazil are huffing, HFOO, and PEP, followed by volume-based techniques. There were regional differences in therapeutic recommendations found between centers and in function of the age of the patient, but not based on the severity of pulmonary function. The knowledge of the devices and techniques currently used for respiratory physical therapy may serve as the base for the search of a consensus in the physical therapeutic treatment of patients with CF.

Conflicts of interest

The authors have no conflicts of interest to disclose.

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