European Journal of Pediatrics (2022) 181:2961–2970
https://doi.org/10.1007/s00431-022-04509-5

ORIGINAL ARTICLE

Does the effect of comprehensive respiratory physiotherapy home-program differ in children with cystic fibrosis and non-cystic fibrosis bronchiectasis?

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Received: 17 February 2022 / Revised: 12 May 2022 / Accepted: 14 May 2022 / Published online: 20 May 2022

Abstract

Bronchiectasis is a form of airway damage as a consequence of endobronchial infection and inflammation and may be present in different diseases. The underlying aetiologies include both cystic fibrosis (CF) and a group of non-cystic fibrosis diseases (NCFB) such as immunodeficiency, primary ciliary dyskinesia, or severe pulmonary infection. Although children with CF and non-cystic fibrosis bronchiectasis (NCFB) have many similar clinical features, their responses to exercise may be different. The aim of this study was to compare the efficacy of a comprehensive respiratory physiotherapy (CRP) home-program in children with CF and NCFB. Thirty children with CF and thirty children with NCFB were included in the study. Both groups performed the CRP home-program twice daily for 8 weeks. Pulmonary function, exercise capacity, and respiratory and peripheral muscle strength were assessed at baseline and after 8 weeks of training. Both groups experienced significant improvements in pulmonary function, exercise capacity, and respiratory and peripheral muscle strength ($p < 0.001$). Maximum expiratory pressure, exercise capacity, and peripheral muscle strength were further improved in NCFB group compared to CF ($p < 0.05$); however, there was a great variability in the improvements for each variable.

Conclusion: CRP is beneficial both for children with CF and NCFB and adherence to the program was high in both groups.

What is Known:
- *Different physiotherapy approaches in the management of non-cystic fibrosis bronchiectasis have been based on the experience gained from the research studies performed in cystic fibrosis.*
- *Although having similar pathophysiology, these two diseases show variation in some pulmonary and extrapulmonary features.*

What is New:
- *The respiratory muscle strength and the efficacy of comprehensive respiratory physiotherapy have been compared for the first time in children with cystic fibrosis and non-cystic fibrosis bronchiectasis.*
- *Comprehensive respiratory physiotherapy provides higher increases in children with non-cystic fibrosis bronchiectasis in exercise capacity and expiratory and peripheral muscle strength; however, there was a great variability in these improvements. Nevertheless, it can be concluded that both groups significantly benefited from the CRP program.*

Keywords Cystic fibrosis · Non-cystic fibrosis bronchiectasis · Respiratory physiotherapy · Pulmonary function · Exercise capacity · Respiratory and peripheral muscle strength

Abbreviations

ATS	American Thoracic Society
CF	Cystic fibrosis
CRP	Comprehensive respiratory physiotherapy
FEV_1	Forced expiratory volume in one second
FVC	Forced vital capacity
MEP	Maximal expiratory pressure
MIP	Maximal inspiratory pressure
NCFB	Non-cystic fibrosis bronchiectasis
OPEP	Oscillatory positive expiratory pressure
PEF	Peak expiratory flow
6MWT	6-Minute walk test

Communicated by Peter de Winter

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Introduction

Although children with cystic fibrosis (CF) and non-cystic fibrosis bronchiectasis (NCFB) have similar features of pathophysiology, they show variations in some outcomes of lung diseases [1, 2] and pulmonary and extrapulmonary features [3]. The presence of a genetic deterioration and primary multiple organ failure, malnutrition, and systemic effect in children with CF may affect muscle metabolism negatively, while the reason for the deterioration of muscle metabolism in children with NCFB may be secondary to lung damage [4]. In addition, there were findings of decreased oxygen delivery and desaturation in the patients with CF compared to the patients with NCFB in response to exercise [5]. Therefore, even if the same physiotherapy approaches are applied, the results may differ between the patients with CF and NCFB.

It is stated in the current guidelines that several physiotherapy approaches play an effective role in the management of both diseases [6, 7]. Comprehensive respiratory physiotherapy (CRP) home-program is one of these approaches and includes breathing and thoracic expansion exercises, postural drainage techniques, use of oscillatory devices, and some recommendations related to physical activity. A systematic review has shown that physiotherapy approaches that consist of similar one or more treatment modalities, such as chest physiotherapy or airway clearance strategies, improve pulmonary function, respiratory and peripheral muscle strength, and exercise capacity in patients with CF [8]. On the other hand, a randomized crossover study [9] and a recent review [10] have demonstrated the positive effects of these techniques also in patients with NCFB. In contrast, another review concluded that therapies for patients with NCFB cannot be extrapolated from CF clinical trials [11]. So, there are conflicting reports in the literature on whether similar physiotherapy approaches should be implemented in both CF and NCFB. In addition, how the optimum physiotherapy approach should be is not certain for neither patients with CF nor patients with NCFB.

When the literature emphasizing the importance of patient-oriented different physiotherapy approaches in the management of these diseases is reviewed [8, 10], there seems to be no study with respect to the comparison of the efficacy of CRP home-program in children with CF and NCFB. This study aims to answer this question by exploring the efficacy of a CRP home-program in children with CF and NCFB and compare the outcomes of this program between these two patient groups.

Material and methods

We recruited eligible children with CF and NCFB admitted to the Division of Pediatric Chest Diseases at the Bezmialem Vakif University Hospital who were than referred to the Department of Cardiopulmonary Physiotherapy and Rehabilitation for respiratory physiotherapy (Fig. 1).

Inclusion criteria were being 6–18 years old and clinical diagnosis of CF or NCFB. Exclusion criteria were active participation in any other CRP program; previous participation in supervised, multi-faceted CRP programs; having a previous history of lung or liver transplantation; presence of an acute exacerbation in the last month and/or having a history of hospitalization; and having a diagnosis of orthopedic problems affecting mobility or a history of musculoskeletal surgery.

We estimated that a sample size of at least 20 patients for each group would have a power of 80% to detect a minimum clinically significant difference of 54 m [12] of the 6-min walk test (6MWT) distance which has the highest standard deviation among all variables, with an alpha value of 0.05. Participants were included in the study by calculating the 20% increase in sample size adjusting for the drop-out rate.

Study design

A comparative, open label interventional study was conducted. Pulmonary function (spirometry), exercise capacity, and respiratory and peripheral muscle strength of all subjects were evaluated before and after the CRP home-program by a cardiopulmonary physiotherapist. After the baseline assessments, all subjects performed the CRP home-program twice daily for 8 weeks. The first session was supervised by another cardiopulmonary physiotherapist working in the department, and the rest of the sessions were performed as a home-based exercise program. Children were instructed to keep a diary for home sessions to improve adherence to exercise program, and it was controlled at every supervised session. Adherence (%) was defined as the ratio of the completed sessions to total sessions, which was calculated as "(completed sessions) / (total sessions = 112 sessions) multiplied by 100."

Outcome measures

Pulmonary function

Spirometry was performed with a spirometer (Pony FX; COSMED, Italy) according to the guideline of American Thoracic Society (ATS) and European Respiratory Society [13]. The reference equations were used for predicted values of forced vital capacity (FVC), forced expiratory volume in one second (FEV_1), and peak expiratory flow (PEF) [14].

Exercise capacity

The 6MWT was performed in accordance with the guideline of ATS [15]. The subjects were requested to walk along a

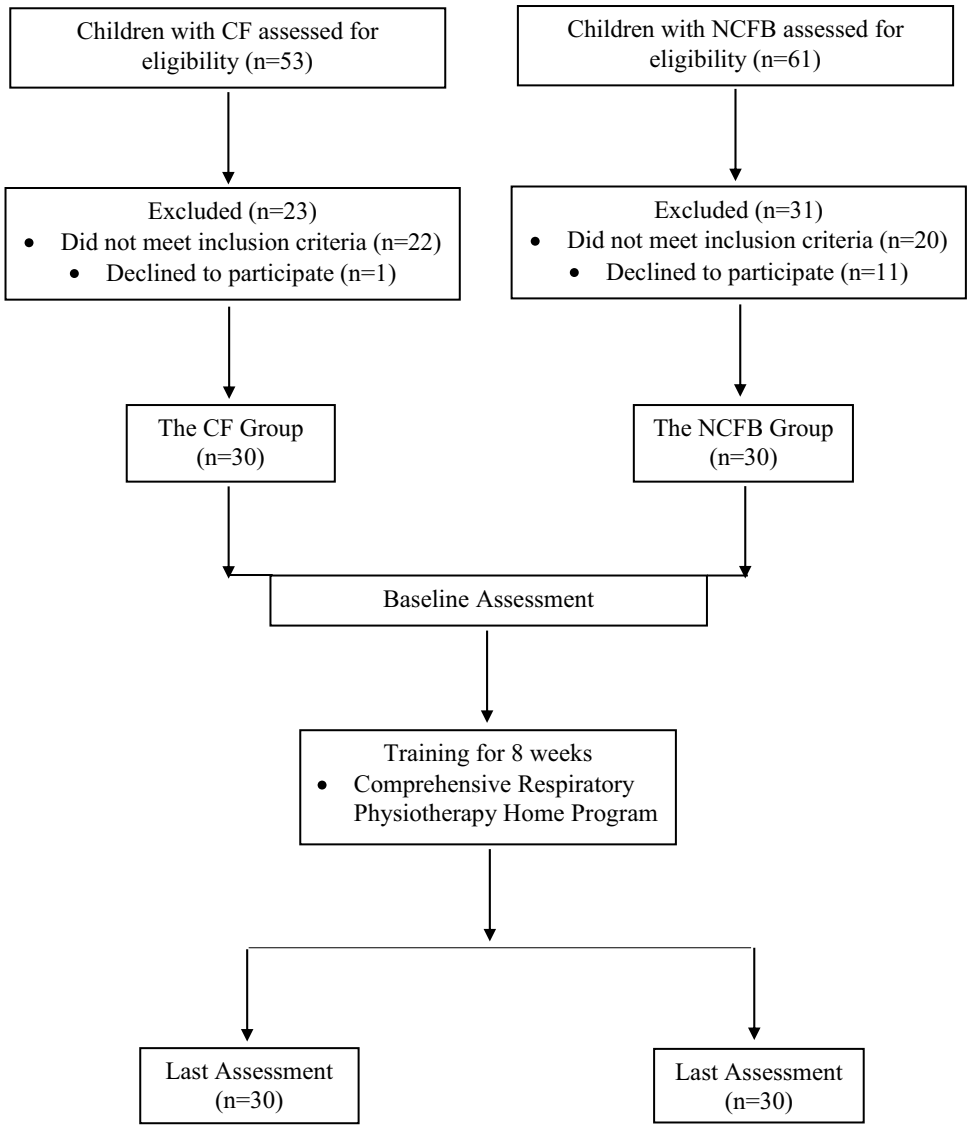

Fig. 1 Flowchart of the study

30-m flat corridor as fast as they could in their walking speed for 6 min. The walking distance in 6 min was recorded. Also, the 6MWT distance was expressed as percentage of predicted values calculated from age, height, and sex [16]. Oxygen saturation and heart rate were also measured before and after the test using a pulse oximeter (Beuer oximeter; Beurer GmbH; Germany). Dyspnea was rated on the Modified Borg Scale. The anchors were "0" for no dyspnea and "10" for maximum dyspnea [17].

Respiratory muscle strength

A portable electronic mouth pressure device (MicroRPM; MicroMedical, UK) was used. Maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP) measurements were recorded. A maximum value of three efforts with alterations less than 10% or 10 cmH2O was accepted as MIP and MEP [18]. The subjects had a rest for 1 min between each test.

Peripheral muscle strength

M. quadriceps was measured using a MicroFet 2 hand-held dynamometer (Hogan Health Industries, USA). Test was repeated three times for the dominant side and the average value in kilograms (kg) was accepted as peripheral muscle strength value of the subject [19]. Subjects were allowed to rest for 1 min between efforts.

Interventions

CRP home-program consisted of controlled breathing technique, diaphragmatic breathing exercises, thoracic expansion

exercises, incentive spirometer (Triflo), oscillatory positive expiratory pressure (OPEP) device (Flutter or Shaker), postural drainage with percussions, coughing techniques, and physical activity counseling. All exercises, including the ones with devices, were carried out in two sets of 5 repetitions with rest intervals of 5–6 tidal breaths between the exercises to prevent respiratory muscle fatigue and hyperventilation. The duration of one session was 40 min which consisted of 20 min of postural drainage and 20 min of other interventions. Postural drainage with percussion was performed in 45° Trendelenburg position during the patient lying on the back, front, left, and right sides for 3–5 min, respectively. Additionally, all the subjects were recommended to perform at least 60 min of moderate-intensity physical activity such as walking, sports, ball games, or simple exercises on Swiss ball or trampoline daily. Moderate-intensity was described as "intense enough to make the child feel like he/she is breathing somewhat hard but can still carry on a conversation" [20].

Statistical analysis

Data were analyzed using the statistical software package SPSS version 20.0 (SPSS Inc., USA) and tested using the Kolmogorov–Smirnov calculation for normality. Paired Sample *T*-test or Wilcoxon Test was used for in-group comparisons. Independent Samples *T*-test or Mann Whitney *U* test was used for between-group comparisons depending on the distribution properties of the data. Categorical variables were compared between groups using chi-square test. Cohen's d effect sizes were calculated for between group differences. The results were considered significant with p values < 0.05.

Results

A total of 114 children were screened for eligibility and 54 of them were excluded (Fig. 1). The final analysis consisted of 60 participants, including 30 children with CF (10.92 ± 2.06 years) and 30 children with NCFB (12.27 ± 3.22 years). Demographics and clinical characteristics of participants are shown in Table 1. The etiologies of bronchiectasis in the NCFB group were detected as primary ciliary dyskinesia ($n = 11$; 36.6%), post-infectious ($n = 8$; 26.6%), idiopathic ($n = 7$; 23.3%), and immunodeficiency ($n = 4$; 13.3%). Nine (30%) children in NCFB group and 30 (100%) children in CF group were routinely receiving inhaled therapy ($p < 0.001$). Regarding former physiotherapy practices, 12 (40%) patients in CF groups were routinely performing at least 1 physiotherapy technique for airway clearance, whereas only 5 (17%) children in NCFB group were routinely performing physiotherapy. Participation in former routine physiotherapy was significantly higher in CF group ($\chi 2 = 4.022$; $p = 0.044$). Adherence (%) to CRP program was 78 ± 22 in NCFB group and 84 ± 16 in CF group, with no between-group difference ($p = 0.233$).

CF was diagnosed based on an abnormal sweat test in all 30 children with genetic confirmation in 28. F508del homozygous ($n = 7$), F508del heterozygous ($n = 5$), N1303K homozygous ($n = 2$), E92K homozygous ($n = 2$), E92K heterozygous ($n = 2$), R347P homozygous ($n = 1$), G542X homozygous ($n = 1$), G85E homozygous ($n = 1$), R1070Q homozygous ($n = 1$), 2183AA->G homozygous ($n = 1$), 1531C/T heterozygous ($n = 1$), 621 + 1G->; T homozygous ($n = 1$), 3755delG homozygous ($n = 1$), and R334W heterozygous ($n = 1$) mutations were identified in gene analysis.

The baseline pulmonary function, exercise capacity, and peripheral muscle strength were not different between groups, except the MEP value ($p = 0.002$). Children with CF had higher baseline MEP value. The baseline pre-treatment and post-treatment values and the effects of the CRP home-program on pulmonary function, exercise capacity, and respiratory and peripheral muscle strength are presented in Table 2. Both groups experienced significant improvements in FVC (%pred), FEV1 (%pred), and PEF (%pred) values ($p < 0.001$) with no differences between the groups. In terms of respiratory muscle strength, there was significant increase in MIP and MEP values in both groups ($p < 0.001$). However, the increase in MEP value was significantly higher in the NCFB group compared to the CF group, having a medium effect size for the difference ($p = 0.037$; $d_{cohen} = 0.54$). Exercise capacity indicated by both 6MWT distance and 6MWT% significantly improved in both CF and NCFB groups ($p < 0.001$), but the magnitude of the improvement in 6MWT and 6MWT% was significantly greater in NCFB group, having a medium effect size for the difference ($p = 0.027$, $d_{cohen} = 0.69$; $p = 0.039$, $d_{cohen} = 0.55$, respectively). Regarding the peripheral muscle strength, there was a significant increase in both groups ($p < 0.001$). The enhancement in quadriceps muscle strength of the NCFB group was statistically higher than the CF group with large effect size ($p < 0.001$; $d_{cohen} = 1.16$).

Variation of the improvements (Δ values) in each variable are present as scatter-plots for each group (Fig. 2). Although the improvements in exercise capacity and expiratory and peripheral muscle strength were significantly higher in the NCFB group compared to the CF group, Δ values in each variable showed great variability. In-group improvements did not seem to have a significant clustering or heterogeneity.

Discussion

This study showed that a multi-faceted CRP home-program provides significant improvements in pulmonary function, exercise capacity, and respiratory and peripheral muscle strength in both children with CF and NCFB. These

Table 1 Demographics and clinical characteristics of participants

	Non-cystic fibrosis bronchiectasis ($n=30$)	Cystic fibrosis ($n=30$)	p value
Age (years)	12.27 ± 3.22	10.92 ± 2.06	0.059
Gender			
Boys	16 (53.3%)	15 (50%)	0.800
Girls	14 (46.6%)	15 (50%)	
Body mass index (kg/m^2)	17.99 ± 2.74	17.60 ± 2.82	0.460
Body mass index (z-scores)	−0.48 ± 1.28	−0.26 ± 0.83	0.066
Age at diagnosis (month)	64.6 ± 18.4	4.5 ± 2.3	**< 0.001**
No. of bronchiectatic lobes (n)			
1	7 (23.3%)	8 (26.6%)	0.619
2	10 (33.3%)	11 (36.6%)	
3 or more	13 (43.3%)	11 (36.6%)	
Pseudomonas aeruginosa colonization	4 (13.3%)	11 (37%)	**0.038**
Routine inhaled therapy	9 (30%)	30 (100%)	**< 0.001**
Former physiotherapy practice			
Routinely performs at least one technique	5 (17%)	12 (40%)	**0.044**
No routine physiotherapy	25 (83%)	18 (60%)	
Formerly practiced physiotherapy technique			
OPEP device/incentive spirometer	4 (13%)	9 (30%)	0.947
Postural drainage	2 (7%)	6 (20%)	
Breathing exercises	1 (3%)	2 (7%)	

Data are presented as mean ± standard deviation or n (%)

improvements had medium and large effect sizes, indicating that CRP can provide clinically significant changes for these patients. The improvements in MEP, 6MWT, and quadriceps muscle strength were higher in the NCFB group compared to CF group. However, because of the presence of a great variability in these improvements, it was not possible to demonstrate a clinical superiority of this increase in one group over another. In addition, the lack of studies showing the minimal clinical important difference in these variables in children with CF and NCFB caused us to be unable to comment on clinical relevance. Although the CRP program required considerable time commitment, adherence to program was rather high and there was no difference between groups in terms of adherence. In addition, time investment in the routine inhaled therapy did not seem to affect adherence to the program either. We believe that multi-faceted, home-based CRP program is feasible for both children with NFCB and CF.

In the literature, effects of chest physiotherapy or airway clearance techniques in CF or NCFB were generally investigated by comparing the effects of one technique to another. Combined effects of the different techniques or whether they provide further improvements for these patients are less studied. Studies including oscillatory devices [8] or various breathing exercises [21] in CF report that such techniques are beneficial for patients, and no technique is superior to another. However, in terms of spirometric variables, these techniques fail to provide statistically significant improvements. Similar findings are reported for NCFB as well, indicating physiotherapy techniques that are applied alone fail to improve spirometry [9, 10]. On the other hand, combined physiotherapy approaches may provide further improvements for spirometric variables in these patients. In our previous study, we showed that home-based CRP improves FEV1 and FVC up to 4% [22]. In our current study, children with CF and NCFB achieved similar and significant improvements in spirometry, indicating that both patient groups may equally benefit from a comprehensive and combined physiotherapy program. Literature suggests that improvements in the spirometric variables may be explained by the mechanical effect resulting from the mucus that has been removed from the airways via physiotherapy techniques [23]. And, considering the improvements in spirometry are similar in both patient groups in the current study, we presume that CRP provided similar mucus transport despite more viscous mucus is present in CF patients. To our best knowledge, there is no study that compares the efficacy of such physiotherapy programs between CF and NCFB. Consequently, we were unable to compare our results and speculate on our assumptions. In the literature, studies on respiratory muscle strength mostly focus on respiratory muscle training, consequently the effect of one or more chest physiotherapy approaches on respiratory muscle strength has not been adequately investigated. Venturelli et al. showed that breathing techniques provide

Table 2 Pre-treatment and post-treatment in-group and between group comparisons

	Non-cystic fibrosis bronchiectasis ($n=30$)				Cystic fibrosis ($n=30$)				Between groups difference delta (Δ)	Effect size
	Pre-treatment	Post-treatment	In-group change (Δ)	*p* value	Pre-treatment	Post-treatment	In-group change (Δ)	*p* value	*p* value	Cohen's d
FVC (% pred)	80.57 ± 12.58	84.87 ± 13.84	4.30 ± 5.28	**< 0.001**	88.13 ± 13.11	91.35 ± 15.51	3.23 ± 3.36	**< 0.001**	0.795	0.24
FEV_1 (% pred)	72.20 ± 16.16	77.50 ± 16.22	5.30 ± 6.85	**< 0.001**	78.36 ± 14.31	82.06 ± 13.53	3.73 ± 4.45	**< 0.001**	0.128	0.27
PEF (% pred)	67.50 ± 17.72	77.50 ± 17.96	10.0 ± 12.12	**< 0.001**	75.91 ± 17.93	89.28 ± 17.56	13.40 ± 11.47	**< 0.001**	0.332	0.28
6MWT distance (m)	570.54 ± 82.41	633.90 ± 66.72	63.35 ± 38.65	**< 0.001**	550.94 ± 56.05	592.67 ± 63.66	40.90 ± 24.18	**< 0.001**	**0.027**	**0.69**
6MWT%	84.46 ± 10.07	93.63 ± 8.31	9.17 ± 5.82	**< 0.001**	85.37 ± 7.00	91.85 ± 8.05	6.47 ± 3.90	**< 0.001**	**0.039**	**0.55**
MIP (cmH_2O)	73.83 ± 17.30	90.04 ± 19.54	16.21 ± 8.24	**< 0.001**	66.84 ± 15.14	87.46 ± 14.84	20.62 ± 11.31	**< 0.001**	0.078	0.44
MEP (cmH_2O)	73.67 ± 25.51	84.33 ± 28.77	10.67 ± 8.19	**< 0.001**	87.65 ± 13.63	94.12 ± 16.04	6.50 ± 7.05	**< 0.001**	**0.037**	**0.54**
Quadriceps muscle strength (kg)	28.42 ± 8.78	34.3 ± 9.93	6.42 ± 3.36	**< 0.001**	31.55 ± 9.67	34.21 ± 10.34	2.69 ± 3.04	**< 0.001**	**< 0.001**	**1.16**

Data are reported as mean ± standard deviation

FVC forced vital capacity, *FEV1* forced expiratory flow in 1 s, *PEF* peak expiratory flow, *pred* predicted, *6MWT* 6-min walk test, *m* meter, *MIP* maximum inspiratory pressure, *MEP* maximum expiratory pressure

an increase in MIP and MEP values in patients with chronic respiratory diseases [24]. In our previous study, we also found that a CRP program provided a significant increase in MIP and MEP values in children with CF [22]. Since the inspiratory muscles are morphologically and functionally skeletal muscle, they will respond to training in a similar way as any skeletal muscle when the appropriate physiological load is applied [25]. Consequently, it is not surprising to achieve increments in MIP value by implementing breathing exercises that involve repetitive maximal inspiration maneuvers. Also, Paiva et al. reported that incentive spirometry can provide a significant increase in MIP value by creating high-intensity muscle activity, since it encourages individuals to perform higher volumes of breathing and enables the inspiratory muscles to contract with higher strengths with contractility [26]. Both the combination of several breathing techniques in the CRP program and the standard incentive spirometry training given to both groups may have played a role in the similar increase in MIP value in both groups. Regarding expiratory muscles, there was also a significant increase in MEP value in both groups; however, the increase was statistically higher with great variability of MEP Δ value in the NCFB group compared to the CF group. Also, baseline MEP value significantly was higher in the CF group compared to the NCFB group in our study. The higher increase obtained in the NCFB group compared to the CF group may be related to the lower baseline value in the NCFB group. Based on studies on mucus properties and cough frequency [1, 27], this result may be relevant for the frequent need for coughing against purulent and thick mucus in patients with CF which may have increased expiratory pressure by creating a pseudo-exercise effect. In addition, the higher rate of participation in former physiotherapy practice in children with CF may have been effective in the higher baseline MEP value. There are studies showing that the use of OPEP devices significantly increases the MEP value with forced and long expiration mechanism in patients with NCFB [9, 28]. In our study, the reasons for the significant increase after treatment in both groups may be the OPEP devices routinely used in the CRP program and the strengthening of the expiratory respiratory muscles with repetitive exercises similar to skeletal muscles [25].

In recent years, there has been a lot of research on exercise capacity due to its relation with physical activity, quality of life, morbidity, and mortality [5]. Although there is no clear opinion about the reasons for decreased exercise capacity in children with CF and NCFB, the common finding is that a multifactorial cause including decrease in respiratory and peripheral muscle strength, impairment in respiratory function and oxygen delivery, and nutritional deficits [5, 29]. In the present study, the CRP home-program produced significant changes in both groups; however, the improvement in the NCFB group was significantly higher than in the CF group.

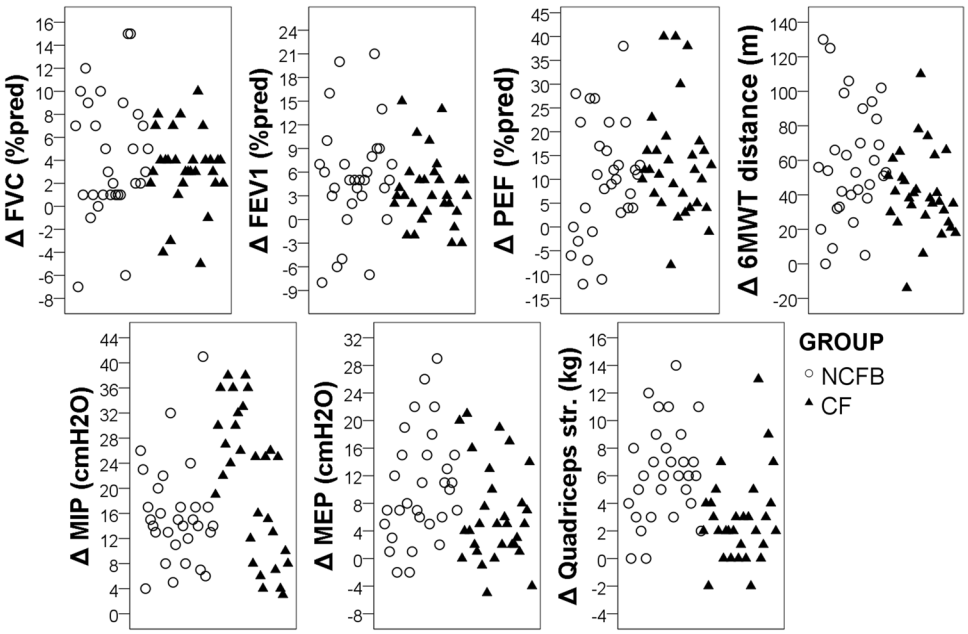

Fig. 2 Variations of the improvements (Δ values) in each variable

Although 6MWT further improved in NCFB group, it should be carefully interpreted since the improvements showed great variability. It has been reported that 6MWT has a positive relationship with pulmonary function [30, 31] and respiratory muscle strength [32–34] in patients with both NCFB and CF. In addition, Hebestreit et al. reports that exercise capacity is associated with FEV1 (%pred) and muscle strength in patients with CF as well [35]. Accordingly, improvement of exercise capacity in our study may be attributed to the increases in pulmonary function and respiratory and peripheral muscle strength. However, the increase in 6MWT was statistically higher in children with NCFB compared to children with CF group. The fact that the NCFB group reached significantly higher values of MEP and quadriceps muscle strength may explain the augmented and significant improvement in this group compared to the CF group. Vendrusculo et al. stated that peripheral muscle strength is associated with aerobic fitness in children with CF [36]. Similarly, Troosters et al. identified quadriceps weakness in adult patients with CF and found it to be associated with functional exercise capacity and time spent in physical activity of moderate intensity [34]. Supporting this, there are studies showing that aerobic exercise increases peripheral muscle strength in both CF and NCFB patients [37, 38]. Moderate-intensity physical activity counseling was part of the CRP program in our study. The high adherence to physical activity recommendations of the children in both groups may explain the increase in peripheral muscle strength. In the present study, the quadriceps muscle strength further improved in the NCFB group compared to the CF group; however, there was a great variability in this improvement. Since children with CF are diagnosed at an earlier age and start their routine treatments earlier, they are more familiar with physical activity, and this may have led to higher baseline quadriceps muscle strength values. Therefore, children with CF may not have benefited from the CRP program in terms of peripheral muscle strength as much as children with NCFB.

Most of CF patients receive time-consuming nebulizer treatments such as dornase alfa, hydrator, or antibiotic therapy for the maintenance of lung health [39]. NCFB patients may also receive nebulizer treatments such as bronchodilators, hydrators, or antibiotics [40]; however, they are not as common and intense as that in CF patients. It may be speculated that time commitment for such treatments may interfere with the time that patients are willing to invest in physiotherapy practices. However, CPR adherence was not significantly different between groups in our study. Accordingly, we believe that time investment in inhaled therapies does not affect the commitment to the physiotherapy program in these patient populations. Although adherence is similar in groups, it is important to highlight that declining ratio for participation in the study was much higher in children with NCFB. This may be due to the fact that children with NCFB do not have as severe and symptomatic disease as children with CF, and, consequently, they may not feel the need to receive additional treatment. This assumption is supported by the study of Basaravaj et al. that reports NCFB patients with more severe disease or hospitalizations or frequent exacerbations perform airway clearance techniques more often [41]. In addition, CF patients are diagnosed at an earlier ages and start their routine treatments earlier. Consequently, these patients become more familiar with additional rehabilitative approaches. We believe this may have also provided higher participation rate for children with CF.

Major strength of our study was that the effects of a multifaceted and CRP program were compared between CF and NCFB for the first time. Despite reporting original findings with a clinical significance, this study has some limitations. The lack of a control group and investigation of the efficacy of the CRP home-program on etiological conditions and comorbidities might limit the findings of our study. Furthermore, we could not consistently collect any data concerning the bacterial colonization of the bronchiectasis. Finally, we were not able to measure sputum wet weight on a regular basis since the CRP home-program was mainly applied at home by children's parents.

Conclusion

A CRP home-program including breathing exercises, incentive spirometer and OPEP devices, postural drainage, and physical activity counseling provided significant improvements in pulmonary function, exercise capacity, and respiratory and peripheral muscle strength in both the children with CF and NCFB. Although improvements in exercise capacity and expiratory and peripheral muscle strength were higher in the NCFB group, there was a great variation in these improvements. Nevertheless, we may conclude that both groups significantly benefited from the CRP program. On the other hand, CRP program was feasible, and adherence was high in both groups. Our results suggest that inclusion of CRP home program in the general management of the children with CF and NCFB may provide further benefits in many pulmonary and extrapulmonary features.

Authors' contributions HNG: designed and supervised the study, analyzed and interpreted the data, and critically reviewed the manuscript. HU: involved in designing the study, performed the assessments, collected the data, performed statistical analysis, and drafted the manuscript. MZ: involved in designing the study, implemented the training of the children, and interpreted the statistical analysis. HDK: involved in designing the study, implemented the training of the children, and reviewed the literature. EC: involved in designing the study and recruitment of the children and interpreted clinical data. All authors contributed to the final version of manuscript and approved it to be published.

Availability of data and materials Not applicable.

Code availability Not applicable.

Declarations

Ethics approval The study was approved by the ethics committee of Bezmialem Vakif University (protocol number: 18/351) and registered to ClinicalTrials.gov website (registration number: NCT04170114). The study was conducted in accordance with the Declaration of Helsinki.

Consent to participate Informed consent was obtained from parents of all participants included in the study.

Consent for publication All parents of participants gave informed consent to publish data from the study.

Conflict of interest The authors declare no competing interests.

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