Physical therapy

Chest physical therapy in cystic fibrosis

Code: 052 **Updated:** April 29, 2024

Background

Chest physical therapy (CPT) is an essential component of chronic maintenance therapy (Castellani C et al, 2018) in CF, with the aim to clear the airways of mucus secretions, but it's time consuming.

Since the life expectancy in CF has increased over the past decades with a corresponding slower rate of decline in pulmonary function newer airway clearance techniques, such as active cycle of breathing and autogenic drainage, have been demonstrated to be as effective as conventional chest physiotherapy offering many advantages. Techniques such as PEP mask therapy, high pressure PEP mask therapy, airway oscillating devices, and external high frequency chest compression can be used independently of an assistant, thus offering many advantages with a more flexible approach to airway clearance management. However, it has not still designed as the best method of airway clearance for patients with CF. Interaction between lung mechanics and the state of the individuals lung disease plays an important role in how a person with CF responds to CPT.

In most cases there is little evidence to support the use of one technique over another. Exercise and physical activity should be integral to the overall physiotherapy management suggested for every individual with CF, irrespective of age and disease severity.

Issues

There is limited evidence to quantitatively assess the effectiveness of CPT in the treatment of patients with CF. A lack of knowledge is registered as to:

1. when it should be commenced;
2. whether there are risks associated with performing different techniques;
3. what is the evidence to support the use of one method to another;
4. which outcome measures are the most appropriate to use in large clinical trials.

What is known

One CDSR (Jones M, 2023) investigated interventions for improving adherence to airway clearance treatment and exercise in people with cystic fibrosis. Two RCTs (77 participants with CF; age range 2 to 20 years; 44 (57%) males) met the inclusion criteria of this review. One study employed an intervention to improve adherence to exercise and the second an intervention to improve adherence to ACT. Both studies measured outcomes at baseline and at three months, but neither study formally assessed the primary outcome of adherence and results were dependent on self?reported data. All evidence was graded as very low certainty. Authors are uncertain whether a music?based motivational intervention may increase adherence to ACTs or affect the risk of hospitalisation for a respiratory infection. They are also uncertain whether an educational intervention increases adherence to exercise or reduces the frequency of respiratory infection?related hospital admission. However, these results are largely based on self?reported data and the impact of strategies to improve adherence to ACT and exercise in children and adolescents with stable CF remains inconclusive. Given that adherence to ACT and exercise therapy are fundamental to the clinical management of people with CF, there is an urgent need for well?designed, large?scale clinical trials in this area, which should conform to the CONSORT statement for standards of reporting and use appropriate, validated outcome measures. Studies should also ensure full disclosure of data for all important clinical outcomes.

One CDSR (Main E, 2023) compared conventional chest physiotherapy compared to other airway clearance techniques for cystic fibrosis. This review included 21 randomised or quasi-randomised clinical trials (N = 778); studies of less than seven days duration were excluded. Studies were conducted in the USA (10), Canada (five), Australia (two), the UK (two), Denmark (one) and Italy (one) with a median of 23 participants per study (range 13 to 166). Participant ages ranged from newborns to 45 years; most studies only recruited children and young people. Most studies compared modifications of CCPT with a single comparator, but two studies compared three interventions and another compared four interventions. The interventions varied in the duration of treatments, times per day and periods of comparison making meta?analysis challenging. All evidence was very low certainty. Authors concluded that there is uncertainty whether CCPT has a more positive impact on respiratory function, respiratory exacerbations, individual preference, adherence, quality of life, exercise capacity and other outcomes when compared to alternative ACTs as the certainty of the evidence is very low. There was no advantage in respiratory function of CCPT over alternative ACTs, but this may reflect insufficient evidence rather than real equivalence. Narrative reports indicated that participants prefer self?administered ACTs. This review is limited by a paucity of well?designed, adequately powered, long?term studies. This review cannot yet recommend any single ACT above others; physiotherapists and people with CF may wish to try different ACTs until they find an ACT that suits them best.

One CDSR (Warnok L, 2023) included 11 cross?over studies (153 participants) and one parallel study (41 participants). There were differences between studies in how the interventions were delivered. The substantial heterogeneity in the treatment interventions

precluded pooling of data for meta?analysis; all studies were judged at unclear risk of performance bias due to the lack of blindeness. The evidence from this review shows that airway clearance techniques (ACTs) may have short?term effects on increasing mucus transport in people with CF. All included studies had short?term follow?up. The evidence in this review represents the use of airway clearance techniques in a CF population before widespread use of cystic fibrosis transmembrane conductance regulator (CFTR) modulators. Further research is needed to determine the effectiveness and acceptability of airway clearance in those treated with highly effective CFTR modulators.

One CDSR (Patterson DK, 2022) selected randomised controlled studies (RCTs) and quasi-RCTs to compare the effect of exercise to other ACTs for improving respiratory function and other clinical outcomes in people with CF and to assess the potential adverse effects associated with this ACT. Four RCTs included 86 participants with a wide range of disease severity (forced expiratory volume in one second (FEV_1) ranged from 54% to 95%) and age 7 to 41 years old. Two RCTs were cross-over and two were parallel in design. Mainly results did not allow to conclude whether or not exercise is a suitable alternative ACT, and the diverse design of included trials did not allow for meta-analysis of results.

One CDSR (Burnham P, 2021) on autogenic drainage vs all other airway clearance techniques either as a single technique or in combination with other techniques in both pediatric and adult CF patients (n = 212) included 8 RCT and quasi-RCT. There were no statistically significant differences found between any of the techniques used with respect to the outcomes measured except when autogenic drainage was described as being the preferred technique of the participants in one study over postural drainage and percussion. Based on the asssessed studies, autogenic drainage was not found to be superior to any other form of airway clearance technique. Larger studies are required to better evaluate autogenic drainage in comparison to other airway clearance techniques in view of the relatively small number of participants in this review and the complex study designs. The studies recruited a range of participants and were not powered to assess non?inferiority. The varied length and design of the studies made the analysis of pooled data challenging.

One CDSR (Stanford G, 2020) included 10 randomised or quasi-randomised clinical controlled trials comparing different inspiratory muscle training regimens with each other or a control in people with CF. 238 patients of any age, diagnosed by clinical criteria, sweat test or genotyping or both were involved. Primary outcomes were: 1. health-related quality of life - chronic Respiratory Disease Questionnaire (emotion); chronic Respiratory Disease Questionnaire (mastery); 2. pulmonary function tests (performed at rest): i) forced expiratory volume at one second (FEV1) ii) forced vital capacity (FVC) 3. exercise tolerance: i) field-based tests ii) laboratory-based tests. Authors concluded that there is insufficient evidence to suggest whether respiratory muscle training is beneficial or not. Healthcare practitioners should consider the use of this intervention on a case?by?case basis. Further research of reputable methodological quality is needed to determine the effectiveness of respiratory muscle training in people with CF. Researchers should consider the following clinical outcomes in future studies: respiratory muscle function, pulmonary function, exercise capacity, hospital admissions, and health?related quality of life. Sensory?perceptual changes, such as respiratory effort sensation (e.g. rating of perceived breathlessness) and peripheral effort sensation (e.g. rating of perceived exertion) may also help to elucidate mechanisms underpinning the effectiveness of respiratory muscle training.

One CDSR (Oliveira V, 2020) evaluated the physical therapies for postural abnormalities in people with CF who have thoracic kyphosis or scoliosis regardless of age and degree of disease severity. 2 RCTs were included. Interventions consisted in any modality of physical therapy considered relevant for treating postural disorders such as manual therapy (e.g. massage, spinal manipulation, and mobilisation), educational programs, exercise training (individualised or group-based or home-based), Pilates, stretching, GPR, IMT, and yoga. Each intervention of physical therapy has been compared with each other, to no physical therapy, sham treatment or usual care. Authors concluded that due to methodological limitations in the included trials, and in addition to the low quality of the current evidence, there is limited evidence about the benefits of physical therapies on postural abnormalities in people with CF. Therefore, further well?conducted trials with robust methodologies are required considering a prior inclusion criterion to identify the participants who have postural abnormalities.

A CDSR (McIlwaine M, 2019) evaluated the PEP technique for aiways clearance in CF patients. A total of 28 studies (involving 788 children and adults) were included in the review; 18 studies involving 296 participants were cross?over in design. The evidence provided by this review is of variable quality, but suggests that all techniques and devices described may have a place in the clinical treatment of people with CF. Following meta?analyses of the effects of PEP versus other airway clearance techniques on lung function and patient preference, this Cochrane Review demonstrated that there was high?quality evidence that showed a significant reduction in pulmonary exacerbations when PEP using a mask was compared with HFCWO. It is important to note that ACTS should be individualised throughout life according to developmental stages, patient preferences, pulmonary symptoms and lung function. This also applies as conditions vary between baseline function and pulmonary exacerbations.

A CDSR (Irons JY, 2019) included only 1 small RCT (n = 40) undertaken at two paediatric hospitals in Australia. The study evaluated the effects of a singing program on the quality of life and respiratory muscle strength of hospitalised children with CF (mean age 11.6 years, 35% male). This study was limited by a small sample size and a high drop-out rate (21%). There were no significant differences between the groups at either post-intervention or follow up; although by the end of treatment there were some within-group statistically significant increases for both singing and control groups in some of the domains of the quality of life questionnaire Cystic Fibrosis Questionnaire-Revised (e.g. emotional, social and vitality domains). For the respiratory muscle strength indices, maximal expiratory pressure at follow up (six to eight weeks post-intervention) was higher in the singing group, mean difference 25.80 (95% confidence interval 5.94 to 45.66). There was no significant difference between groups for any of the other respiratory function parameters (maximal inspiratory pressure, spirometry) at either post-intervention or follow up. This CDSR concluded that there is insufficient evidence to determine the effects of singing on quality of life or on the respiratory parameters in pwCF. However, there is a growing interest in non-medical treatments for CF and researchers may wish to investigate the impact of this inexpensive therapy on respiratory function and psychosocial well-being further in the future.

1 CDSR (Wilson LM, 2018) included six Cochrane Reviews that compared airway clearance technique, either as a single technique or as a combination of techniques, with no intervention, with coughing, or with another airway clearance technique. The quality of the body of evidence comparing different airway clearance techniques for other outcomes was either low or very low. The authors concluded that patients with CF should choose the airway clearance technique that best meets their needs, after considering comfort, convenience, flexibility, practicality, cost, or some other factor.

1 CDSR (Hilton N, 2018) comparing inspiratory muscle training for CF included 9 studies and showed that this practice varied dramatically with frequency, intensity and duration. The authors concluded that healthcare practitioners should consider the use of respiratory muscle training on a case-by-case basis but further research of reputable methodological quality is also needed.

1 CDSR (Freitas DA, 2018) comparing standard postural drainage (greater (30° to 45° head down tilt) and lesser (15° to 20° head down tilt) vs modified postural drainage (without head down tilt) in infants and young children with CF included no new studies in this update.

One CDSR (Warnok L et al, 2015) summarized the evidence on the effectiveness and safety of various airway clearance techniques in people with CF comparing a form of chest physiotherapy with either no physiotherapy treatment or spontaneous cough alone. Coughing, increased radioactive tracer clearance, amount of expectorated secretions during chest physiotherapy and pulmonary function variables were the main outcomes evaluated in treated groups compared to controls. Among 157 included studies eight cross-over studies (data from 96 participants) met the inclusion criteria for analysis, but with differences between the way that interventions were delivered, with several of the intervention groups combining more than one treatment modality. Results from other studies showed extensive variability regarding the effects of different techniques on different outcomes. Results of this review show that airway clearance techniques have short-term effects in terms of increasing mucus transport. No evidence was found on which to draw conclusions concerning the long-term effects.

A CDSR (McCormack P et al, 2017) evaluated the efficacy of autogenic drainage for airway clearance in CF. Seven studies including 208 patients were eligible for inclusion. One study was a parallel design with the remaining six being cross-over in design; participant numbers ranged from 17 to 75. The total study duration varied between four days and two years. The age of participants ranged between seven and 63 years with a wide range of disease severity reported. There were no statistically significant differences found between any of the techniques used with respect to the outcomes measured except when autogenic drainage was described as being the preferred technique of the participants in one study over postural drainage and percussion.

One RCT (Emirza C, 2021) investigated the effect of Expiratory Muscle Training on Peak Cough Flow (PCF) in Children and Adolescents with CF. 30 CF patients were enrolled. Patients were randomized as training and sham groups. Both groups were trained with the EMT protocol, which involved twice per day for at least five days per week for six weeks. The training intensity in the training group was 30% of the maximal expiratory pressure (MEP). In the sham group, it remained at the lowest pressure (5cmH(2)O). Twenty-eight patients completed the study. Changes in PCF (p=0.041) and MEP (p=0.003) were higher in the training group than the sham group. Also, treatment burden (p=0.008), digestive symptoms (p=0.019), and vitality (p=0.042) in QoL were more improved in the training group. Maximal inspiratory pressure (MIP) (p=0.028) and 6MWD (p=0.035) changed significantly only in the training group. Spirometric measurements did not change (p>0.05). The results of the study show that EMT could improve PCF, MEP, treatment burden, digestive symptoms, and vitality domains of QoL in patients with CF. Moreover, MIP and functional exercise capacity improved in the training group with EMT.

One RCT (Montero-Souza A et al, 2020) evaluated the effect of recorded instrumental music as an adjunct to conventional physiotherapy. A group who perceived specifically composed music was compared to patients that liked music , and to a group with no music during the usual ACT routine in children with CF aged from 2 to 17. Enjoyment and perception of time were evaluated as primary outcome via validated questionnaires. Efficiency was evaluated as secondary outcome. Enjoyment increased after the use of the specifically composed music (children +0.9 units/parents +1.7 units; p<0.05) compared to enjoyment with no music (0 units) and familiar music (+0.5 units), while perception of time was 11.1 min (±3.9) less than the actual time in the treated group (p<0.05). The Authors suggested that the specifically composed, played and compiled instrumental recorded music may be an effective adjunct to ACT to establish a positive response.

One RCT (Dwyer TJ, 2019) investigated the effects of treadmill exercise vs resting breathing and PEP therapy on mucus clearance in 14 adults with mild to severe CF lung disease. Mucus clearance was measured using the radioaerosol technique and gamma camera imaging. Treadmill exercise alone was less effective than PEP therapy (mean difference -7%, 95% CI -6- -8). There were no significant differences in mucus clearance from the intermediate and peripheral lung regions, but significantly less clearance from the central lung region.

One RCT (Zeren M, 2019) investigated the effects of inspiratory muscle training (IMT) on postural stability, pulmonary function and functional capacity in 36 children with CF (age range: 8-18 years) through Biodex Balance system (R), spirometry, respiratory muscle strength and 6-min walk distance (6MWD) at baseline and after 8 weeks of training. Authors' conclusions were that combining IMT with chest physiotherapy (PT) failed to provide further improvements, except for maximum inspiratory pressure (MIP), suggesting that a comprehensive chest PT program may be individually effective in improving overall limits of stability (LOS) score, spirometry, respiratory muscle strength and 6MWD.

One RCT (Vendrusculo FM, 2019) evaluated in CF patients aged 9 years and above if airway clearance physiotherapy (ACT) improves ventilatory dynamics during exercise. 12 CF patients were included in the study with a mean (SD) age of 12.83 (1.85) years. No significant difference in peak oxygen uptake (VO2) was found between the tests. However, lower minute ventilation (VE) and ventilatory equivalents (VEVO2 and VEVCO2) at ventilatory threshold (VT) were noted when ACT was undertaken prior to CPET. The mean(SD) VE (L/min) at VT was 26.67 (5.49) vs 28.92 (6.3) (p=0.05), VEVO2 (L/min) at VT was 24.5 (1.75) vs 26.05 (2.5) (p=0.03) and VEVCO2 (L/min) at VT was 26.58 (2.41) vs 27.98 (2.11) (p=0.03). The authors concluded that ACT prior to exercise may lead to improved ventilatory dynamics during exercise in individuals with CF.

One prospective observational study (Wallaert E et al, 2018) was published including the immediate effects of a single autogenic drainage session on ventilatory mechanics in adult subjects with CF. 13 CF adults were enrolled in the physiotherapy group and 11 CF adults in the control group. No significant changes in any parameter were observed in the control group. A single session of AD improved inspiratory airway resistance, except in the distal airways. The forced oscillations technique provides a new tool for understanding the pathophysiological effects of airway clearance physiotherapy in CF.

Recently (Grosse-Onnebrink J et al, 2017) data from a randomized controlled study, including 20 CF patients (7-34 years) hospitalized for infective pulmonary exacerbation, suggested that a single treatment of HFCWO can have a short-term decreasing effect on the lung clearance index, compared to a control group that received no treatment. These results have to be evaluated in trials using LCI as an endpoint, being the timing of CP in relation to Multiple Breath washout as a possible bias.

In 21 paediatric CF patients with normal baseline spirometry a randomized controlled cross-over study (Jana P, 2021) investigated the change assessed by lung clearance index (LCI) in global ventilation inhomogeneity after 30 minutes reflex zone stimulation technique (RST) compared to that of sham therapy. The interventions were performed in random order and planned 6 months apart. After the RST intervention, the LCI(2.5) (p = 0.004) and Scond*Vt (p = 0.009) decreased significantly, while inspiratory capacity increased (p = 0.012). In the sham-therapy group, none of the parameters changed significantly. Trunk deformity was seen in 76.5% of all patients, and 92.9% of those with trunk deformity showed a decrease in LCI(2.5) after RST. RST has multiple positive short-term effects on lung function, especially in CF patients with trunk deformities.

A retrospective review in children with cystic fibrosis (Boonjindasup W, 2021) investigated the effect of singing on respiratory muscle strength in 35 children with CF. On two different days (T1, T2) children performed MIP/MEP with at least ten attempts each to achieve?<?10% repeatability. All children achieved repeatable MIP/MEP values within 10-11 attempts with 24 (68.6%) and 26 (74.3%) of these achieving best values of MIP and MEP, respectively, at attempts 6-11. Median values of the pressures by three, five, eight and all attempts significantly increased with more attempts (all p?<?0.05). At T2, 56% required fewer attempts to achieve best values, but 32% required more attempts, indicating that the number of attempts required was inconsistent between test days. Authors concluded that at least ten attempts (best two within?<?10% variability) are required to achieve best and reliable MIP/MEP in children with CF. A larger sample size in children with CF and various conditions is required to consolidate these findings.

A prospective cross-over single centre trial (Helper N, 2021) investigated the benefits of mechanical insufflator-exsufflator compared to autogenic drainage in adults with cystic fibrosis. 14 CF males and 8 CF females with an average FEV(1) of 54% received autogenic drainage (AD) or mechanical insufflator-exsufflator (MI-E) in a random order. Sputum was collected and weighed immediately after treatment. Subjects performed lung function tests at baseline, 20?minutes after and 1?hour after treatment; additionally, a 2-minute walk test was performed at the end of all lung function tests. Saturation, dyspnea scores while resting and after a 2-minute walk and subjective fatigue were recorded. 36% more sputum was collected following MI-E than AD treatment (P?<?.0001). A significant difference in saturation in response to the 2MWT was noted in both treatments significantly less desaturation was recorded after the 2MWT in the MI-E treatment (P?<?.01). Treatment with the MI-E was more effective for clearing sputum in CF subjects, initial evidence suggests that the MI-E may be successfully incorporated into treatment protocols. Further studies are needed to assess the long-term benefits of MI-E in patients with CF.

One RCT (Güngör S, 2021) investigated the clinical effects of combining postural exercises with chest physiotherapy in 19 pediatric CF patients. Respiratory functions were improved in both groups; however, these changes were not statistically significant. The MST increased after treatment in both groups (p<0.001 and p=0.003 respectively), without a significant difference between the groups. Emotional function and treatment difficulties subdomains in CFQR were significantly increased only in the group with postural exercises (p<0.05). The authors concluded that the postural exercise program in addition to chest physiotherapy in pediatric CF patients whose postural changes were not taken place did not cause significant changes in respiratory function, exercise tolerance, and postural stability; however, it affected the emotional state well and improved the compliance with the treatment.

One RCT (Fettes E, 2022) investigated if pediatric CF home spirometry require physiologist supervision. 61 CF children were randomly allocated to either supervised or unsupervised home spirometry following a detailed training session. Home spirometry was performed every 2 weeks for 12 weeks. Tests were assigned a quality factor (QF) using centre laboratory grading system as per American Thoracic Society/European Respiratory Society standards, with tests marked from A to D, or Fail. In centre laboratory, authirs aim for QF A in all spirometry tests, but report results of QF B or C with a cautionary note. QF A was, therefore, the primary outcome, and QF A-C, the secondary outcome. 166 measurements were obtained in the supervised group, and 153 in the unsupervised group. Significantly more measurements achieved QF A in the supervised compared to unsupervised group (89% vs. 74%; p?=?<0.001), while proportions reaching Grade A-C were similar (99% vs. 95%; p?=?0.1). All significant declines in spirometry results had a clinical rather than technical reason. Family/patient feedback for both arms was very positive. These results suggest that home spirometry in children should ideally be remotely supervised by a physiologist, but acceptable results can be obtained if resources do not allow this, provided that training is delivered and results monitored according to centre specific protocol.

One RCT (Trimble A, 2022) investigated the effect of three different physiotherapy methods to augment cough-clearance in addition to cough-clearance alone (high-frequency chest-wall oscillating vest, oscillatory positive expiratory pressure, and whole-body vibration) in 10 adults with CF. No differences were identified between any method of airway clearance, including cough clearance alone. Changes in certain small molecule concentrations in exhaled breath following airway clearance were identified. Due to the limitations of this study, the authors do not believe the negative results suggest a change in clinical practice with regard to airway clearance. Findings pertaining to small molecules in exhaled breath may serve as future opportunities for study.

One single-center, prospective cross-over study (Vandevoor TB, 2022) investigated the short term influence of chest physiotherapy on lung function parameters in children with CF (n=17) and primary ciliary dyskinesia (n=14). For the whole group, there was no difference in median change of FEV(1) pp between the treatment and the control group (p 0.969), nor in median change of LCI (p 0.294). For the CF subgroup, the mean change in FEV(1) pp with ACT was -1.4% (range -9 to + 5) versus -0.2% (range -6 to + 5) for no ACT (p 0.271), the mean change in LCI with ACT was + 0.10 (range -0.7 to + 1.2) versus + 0.17 (range -0.5 to + 2.8) for no ACT (p 0.814). In the PCD subgroup, the mean change in FEV(1) pp with ACT was + 1.0 (range -7 to + 8) versus -0.3 (range -6 to + 5) for no ACT (p 0.293) and the mean change in LCI with ACT was -0.46 (range -3.7 to + 0.9) versus -0.11 (range -1.4 to + 1.3) for no ACT (p 0.178). There was no difference between PCD and CF for change in FEV(1) pp after ACT (p = 0.208), nor for LCI (p = 0.095). Authors concluded that in this small group of pediatric patients, no significant short-term effect of chest physiotherapy on FEV(1) pp nor LCI in PCD and CF values nor variability was documented.

One N-of-1 RCT (Gursli S, 2022) investigated airway clearance physiotherapy in six CF adults. Each trial included eight weeks of treatment, twice a week, using saline inhalation in horizontal positions, one with SCT and one with FET, in random order. Efficacy was measured by sputum wet weight (g) after each session. Perceived usefulness and preference were self-reported at the end of the study. Lung function was assessed at baseline and at the end of study. HRQoL was measured using the Cystic Fibrosis Questionnaire-Revised (CFQ-R) at baseline (week 1) and at completion of the study (week 8). Patient-reported outcomes were completed by all subjects. Individual CFQ-R-Respiratory Symptoms Scores (CFQ-R-RSS) showed a positive change, meeting the minimal important difference (MID) ? 4 points in five participants and a negative change in one individual. A strong correlation (r = 0.94 (p<0.01) was found between total sputum weight (g) and the positive changes in CFQ-R-RSS, and between changes in lung function

and CFQ-R-RSS (r = 0.84 (p = 0.04). In conclusion the airway clearance intervention was associated with clinically meaningful changes in patient-reported symptoms on the CFQ-R in the majority of the participants. This finding warrants further investigation regarding treatment, duration and frequency. A long-term study may reveal beneficial effects on other clinically meaningful endpoints, such as pulmonary exacerbations, high-resolution computed tomography scores and HRQoL.

One intervention study (NCT04094441) has completed recruitment of 30 CF childrean (4-18 years of age) to evaluate modeling of chest physiotherapy using impedance measurements (PHYSIOMOD). The impedance will be measured before and after chest physiotherapy using the Impulse Oscillometry System. Results are not available.

One RCT (NCT03965832) is completed to evaluate HFNT (high flow nasal therapy) during exercise in 20 adult CF patients. HFNT is a device that provides patients with air or a blend of air and oxygen at flows up to 60 L/min. In CF, HFNT is routinely used for patients admitted with acute respiratory failure (inability to maintain adequate oxygenation) with positive results. This pilot monocentric study has the aim to understand if HFNT can improve the exercise tolerance in patients with CF and advanced lung disease, by reducing breathlessness and avoiding the drop in oxygenation observed during simple training. Results are not available.

One RCT (Flores J, 2023) inestigated the effects of an early rehabilitation program in 34 adult cystic fibrosis patients during hospitalization due to pulmanry exacerbation. The intervention group underwent an early rehabilitation program for 14 days after admission. In the intervention group, there were increases in 1 repetiiton maximum test (= the maximum amount of weight that the patient could lift once during a standardized weightlifting exercise. This test is used to measure muscle strenght) biceps (P=0.009), triceps (P=0.005), shoulder abductors (P=0.002), shoulder flexors (P=0.004), hamstrings (P<0.001), and quadriceps values (P<0.001). In addition, there were improvements in CFQ-R-emotion (P=0.002), treatment burden (P=0.002), vitality (P=0.011), and physical scores (P=0.026), and a reduction in the Borg resting fatigue score (P=0.037). The interleukins levels did not change after the intervention. The authors concluded that in adult CF patients with pulmonary exacerbation, early hospital rehabilitation had a significant impact on improving resting fatigue, muscle strength, and quality of life.

One systematic review (Cai W, 2024) investigated the effect of respiratory muscle training on children and adolescents with cystic fibrosis. Up to July 2023, electronic databases and clinical trial registries were searched. Controlled clinical trials comparing respiratory muscle training with sham intervention or no intervention in children and adolescents with cystic fibrosis. Six studies with a total of 151 participants met the inclusion criteria for this review. Two of the six included studies were published in abstract form only. Four studies were parallel studies and two were cross-over designs. There were significant differences in the methods and quality of the methodology included in the studies. The pooled data showed no difference in respiratory muscle strength, lung function, and exercise capacity between the treatment and control groups. However, subgroup analyses suggest that inspiratory muscle training is beneficial in increasing maximal inspiratory pressure, and qualitative analyses suggest that respiratory muscle training may benefit respiratory muscle endurance without any adverse effects. In conclusion this systematic review and meta-analysis indicate that although the level of evidence indicating the benefits of respiratory muscle training is low, its clinical significance suggests that we further study the methodological quality to determine the effectiveness of training.

One RCT (Sosa-Pedreschi A, 2024) investigated the effects of a remotely supervised resistance training program on muscle strength and body composition in adults with cystic fibrosis (n=23; age 32.13?±?7.72?years). A remotely supervised, individualized 8-week resistance training program of moderate to high intensity. The exercise group (EX) performed three 1-h resistance training sessions per week over 8?weeks. The control group (CON) followed the physical activity recommendations of their physician. The intervention showed a significant beneficial effect on leg press strength, with a large effect size, both in absolute (p?=?0.011; ?p2 ?=?0.281) and relative (p?=?0.007; ?p2 ?=?0.310) terms. Large intervention effects were observed on total fat mass (p?<?0.001; ?p2 ?=?0.415), body adiposity index (p?<?0.001; ?p2 ?=?0.436), and fat mass index (p?<?0.001; ?p2 ?=?0.445), all showing reduction in the EX group. In addition, significant large size effects were detected on total fat-free mass (p?=?0.046; ?p2 ?=?0.177), trunk fat-free mass (p?=?0.039; ?p2 ?=?0.188), and fat-free mass index (p?=?0.048; ?p2 ?=?0.174), all favoring exercise. No significant effects were observed on pulmonary function and quality of life. An 8-week remotely supervised resistance training program, with moderate to high intensity, effectively improved lower limb muscle strength and body composition.

Unresolved questions

An ongoin RCT (NCT03655249) aims to evaluate the effect of autogenic dreinage (AD) on ventilation inhomogeneity (VI) using the LCI technique in children and adults with CF. Data are estimated to be completed within December 2021. No further information is available.

Data from clinical trials suggest that blinding of participants, caregivers or clinicians in airway clearance studies is quite impossible; on the other hand lack of protocol data increased assessment of risk of bias for the majority of studies.

Based on these data it would be helpful to provide guidelines to healthcare professionals on the use of physiotherapy in CF.

The effect of highly effective modulator therapies with ETI treatment and their impact on ACTs and nebulised treatments has not well studied. Based on the opinions of children with CF, their parents/carers and healthcare professionals (HCPs) some concernes remain about the longer-term outcomes of reducing ACTs or nebulised treatments (Almuhlem M et al, 2022).

Other results on this topic are included in the following related topics:

- *Exercise and physical training*
- *Oscillating device*
- *Positive Expiratory pressure*
- *Active cycle of breathing technique.*

Keywords

Airway clearance technique;