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REVIEW ARTICLE

Systematic Review of the Methodological Quality and Outcome Measures Utilized in Exercise Interventions for Individuals with Cystic Fibrosis

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ABSTRACT

Background: The aims of this study were to evaluate the methodological quality of interventional exercise studies in individuals with cystic fibrosis (CF) and link the reported outcome measures of these studies to the International Classification of Functioning, Disability and Health (ICF) model.

Methods: Electronic searches of PubMed, Scopus, Embase, CINAHL, SPORTDiscus, and Cochrane review were performed in the spring of, 2022. The methodological quality of studies was evaluated using the Effective Public Health Practice Project Quality Assessment Tool (EPHPP). Outcome measures were linked to the categories and sub-categories of the ICF model using ICF linking rules.

Results: We screened 4,769 studies and extracted data from 89 eligible manuscripts. Of these. 41% were one group, pre-post design; 36% were RCTs; 11% were two-group pre-post design; 6% were controlled clinical trials; and 6% were either case reports or case series. Using the (EPHPP), 49% were ranked as high quality, 34% moderate, and 17% low. In addition, body function was the most addressed ICF category followed by activities/participation and, subsequently, body structure.

Significance: This review supports the recommendation of exercise as an intervention to improve a variety of outcomes, ranging from Body structure (e.g., body composition) to Participation (e.g., engaging in community, social, and civic life). We recommend that investigators review the spectrum of potential ICF outcomes when designing rehabilitation trials, to both reflect the potential effects of exercise on a variety of outcomes, but also to facilitate mechanistic studies that are able to link which body structures and body functions are most important to improving patient outcomes at the activity and participation domains. Addressing environmental factors within the ICF is also important for people with CF, such as resource allocation towards exercise and rehabilitation resources. Finally, this review highlights the need for more vigorous methodological designs to minimize risk of bias and improve quality of exercise studies in CF.

Keywords: Cystic Fibrosis, exercise, ICF, quality assessment, systematic review.

1. Introduction

Cystic fibrosis is a genetic disorder that affects almost 40,000 individuals in the United States. It impacts the pulmonary, digestive, and reproductive systems[1]. People with CF often develop bronchopneumonia with recurrent exacerbations that can lead to respiratory failure. The common reported symptoms that individuals with cystic fibrosis complain of are shortness of breath, thickened mucus, poor growth, and cough[1,2]. Existing literature also states that people with CF experience skeletal muscle dysfunction which results in exercise intolerance[3-5].

Exercise training has been proven to be beneficial for individuals with cystic fibrosis.[5] Based on the literature, exercise enhances aerobic capacity, muscle function, and quality of life in people with cystic fibrosis. Studies also support the evidence that exercise training in people with CF improves exercise tolerance and increases cardiovascular and pulmonary functions[6,7]. In individuals with CF, the International Classification of Functioning, Disability and Health (ICF) model is beneficial for clinical decision making, including guiding interventions, predicting outcomes, and assessing relations between its categories[8]. However, little evidence exists regarding the significance of ICF in individuals with CF. More importantly, no previous review has evaluated the link between outcome measures in people with CF and the categories of the ICF model in relation to exercise training and related outcomes. Relating outcome measures pertaining to exercise to the ICF model is important, as it can provide manifest evidence for which ICF categories are both most frequently used as well as less frequently targeted. Thus, shedding light on this issue can inform future studies to consider addressing other relevant categories[9].

Methodological quality is another limitation in exercise intervention studies for people with CF. Although there is growing evidence from randomized controlled trials of exercise for people with CF, a Cochrane review ranked the quality of these trials as low to moderate[10]. Another recent review also reported poor methodological quality in exercise designs within the evaluated studies. According to this review, there is a need for conducting robust designs to provide standardized guidelines for exercise in clinical practice for this population. However, this review only investigated the effects of exercise on lung function and quality-of-life in people with CF using telemedicine[11]. Therefore, there is a gap in the literature regarding methodological quality of all types of exercise intervention studies in individuals with CF. Assessing methodological quality of exercise studies will add to the knowledge about these shortcomings and how to address them in future studies. The aims of this systematic review were therefore to 1) link the reported outcome measures of these studies to the ICF model, and 2) examine the methodological quality of interventional exercise studies in individuals with CF.

2. Methods

2.1. STUDY DESIGN

This study was conducted as a systematic review design, which involved multiple steps, as outlined in the 2020 Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) statement[12]. Our protocol was registered with PROSPERO (CRD42023332797)[13].

2.2. SEARCH STRATEGY

We used a medical librarian (MB) to assist us in formulating our search strategy and selection of databases. the following databases for our initial search: PubMed, Scopus, Embase, CINAHL, SPORTDiscus, and Cochrane review (for details refer to PROSPERO CRD42023332797). Articles were imported into Covidence, an online systematic review tool, to facilitate for article screening and full text reviews[14].

2.3. ELIGIBILITY CRITERIA

The aim of the review was to include studies that evaluated the effects of exercise training in individuals with cystic fibrosis. Studies were eligible if the intervention was any type of exercise or physical activity (including exercises to improve

breathing and respiratory muscle strength) performed by people with CF that lasted at least for a week. Within this intervention the frequency, duration, and mode must be specified. The outcomes needed to be assessed before and after the intervention. Studies were excluded if they were: animal studies, non-English texts, abstracts only, and/or in population who have had a lung transplant or were diagnosed with a different pulmonary disease such as non-CF bronchiectasis. Studies were also excluded if the intervention was airway clearance or chest physical therapy, or the outcomes were in the domain of cost, adherence, and qualitative thoughts about intervention.

2.4. DATA EXTRACTION

Two independent reviewers reviewed the selected studies (NZA and JDL). Two independent investigators (NZA and SC) extracted and linked the outcome measures to the ICF categories; any disagreements were resolved by a third party (JDL). Moreover, two independent reviewers extracted data from RCTs using the PEDro scale and from all other intervention studies using the EPHPP and the modified Sackett's levels of evidence (NZA and NTA). In case of any disagreement between the reviewers, consensus was reached by consulting a third reviewer (JDL and HKY).

2.5. THE INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY AND HEALTH (ICF) LINKING

We used the ICF model[15] to categorize the outcome measures of our included studies. The ICF consists of 4 domains (or constructs): 1) body functions, 2), body structures, 3) activities and participation, and 4) environmental factors (Figure 1). There are multiple "chapters" under each of the four domains. These chapters are also diverged into many codes. Each code is combined of letters and numbers. The letters are derived from the four domains (b-body functions, s-body structures, d-activities and participation, and e-environmental factors). The codes are numerically ordered in different levels. The first level begins with one of the four letters followed by one number, which is the designated chapter number. As the number of levels increases, the description of the outcome measures becomes more specific. For instance, 'heart rate' would go under chapter 4 'functions of the cardiovascular system' of the domain 'b-body Functions', thus b4 is the first level. To classify it more specifically, 'heart rate' which is the third level (b4100) would go under 'heart function' which is the second level (b410).

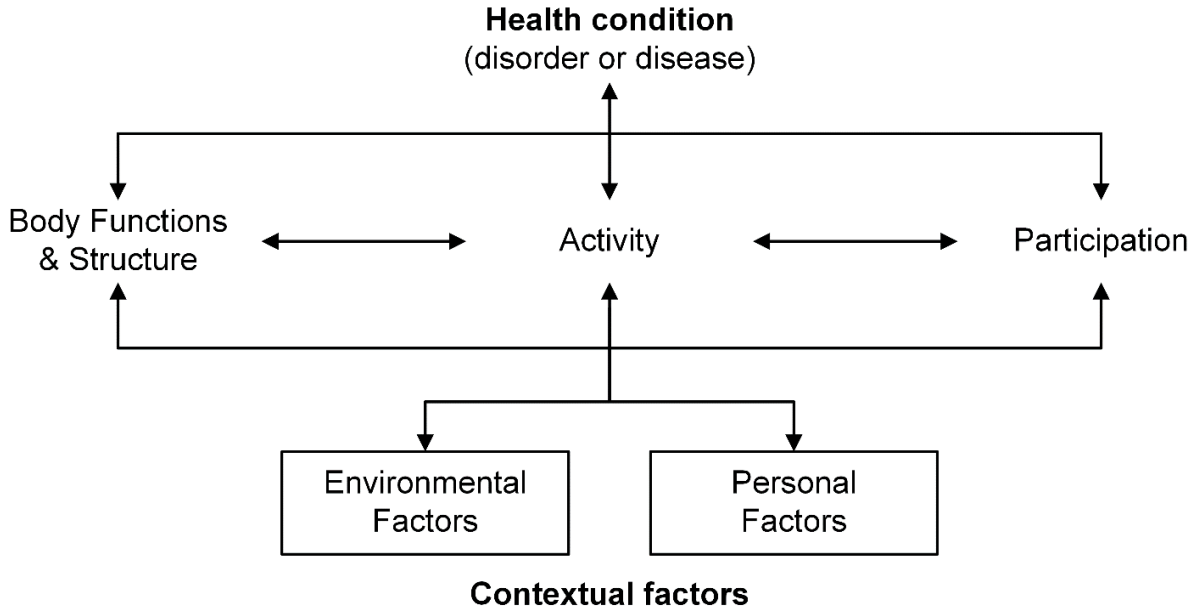

Figure 1. ICF diagram (adapted). Body Functions (b) and Body Structures (s), while included in a single box in the diagram, are distinctly coded "constructs;" Activity & Participation (d), while separate boxes in the diagram, are coded as a single construct; and Environmental Factors (e), one of two Contextual factors, is the only one that is coded.

Linking the outcome measures of this review followed the ICF linking[16-18]. These rules were designed to ensure that each outcome measure is linked appropriately and precisely to one of the ICF categories. If the outcome measures were unidentifiable within the ICF categories or linking any outcome measure was challenging, not definable (nd) or not covered (nc) codes were used for coding.

2.6. METHODOLOGICAL QUALITY AND LEVEL OF EVIDENCE

The Effective Public Healthcare Practice Project "Quality Assessment Tool for Quantitative Studies" (EPHPP)[19] was established and developed by a research team in the school of nursing at McMaster University[20]. This tool is utilized in systematic reviews for the purpose of assessing methodological quality of different types of intervention designs[21,22], and has been previously validated[20,23]. The tool contains six sections, and each section has a rating ranging from 1 to 3 with 1 being strong, 2 moderate, and 3 weak methodological quality[19]. The global quality rating of the six sections depends on the total number of weak ratings. Particularly, no weak ratings indicate a strong quality, one weak rating indicates a moderate quality, and two or more weak ratings indicate a weak quality. Refer for figure 2 for more details about the EPHPP assessment tool[21].

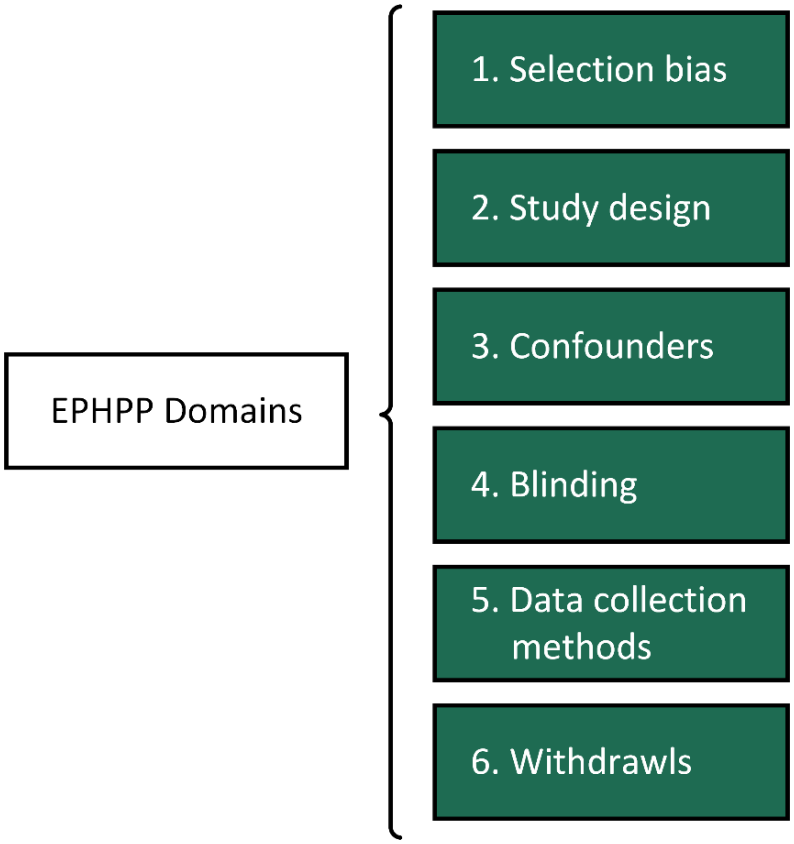

Figure 2. Overview of the EPHPP quality assessment tool domains.

In addition, the Physiotherapy Evidence Database (PEDro) scale was used to evaluate and rate RCT studies[24]. The PEDro scale was reported to be reliable to use in systematic reviews and also appropriate in RCTs of exercise interventions[25]; its validity has also been established[26]. The PEDro scale has 11 items, each item, except the first one, earns a point if the designated item is appropriately addressed. The first item, which asks whether eligibility criteria were specified, only leads the ratings. To be specific, if the answer to the first item was no, then the total score of all items would equal 0. The ratings range from 0 to 10, with 10 being the highest and 0 being the lowest methodological quality.

We also classified the intervention studies using the modified Sackett's levels of evidence which was adopted from Silverman et al.[9] The modified tool was utilized in this review to determine the

strength of evidence of our included studies. Based on this tool, studies were ranked as the following: 1 = RCT (PEDro ≥ 6), 2 = RCT (PEDro score < 6), prospective controlled trial, or cohort, 3 = case control, 4 = one-group pretest–posttest or case series, and 5 = case report, with 1 being the highest and 5 being the lowest level of evidence.

2.7. STATISTICAL ANALYSIS

To illustrate the results of this review, frequency distributions, median analysis as well as simple percentage analysis were performed using Microsoft Excel. Microsoft Excel was also used for Kappa coefficient. Based on Kappa statistics, the agreement between the two reviewers for the EPHPP was 0.88 and for the modified Sackett's levels of evidence was 0.87, both indicating a strong agreement. However, Kappa statistics for the PEDro score was 0.39 indicating a fair agreement, thus, the scoring was taken from the website of the PEDro database[24].

3. Results

We found 4,769 articles in our initial search. The PRISMA flow diagram (Figure 3) illustrates the number of initial studies identified through those finally included.

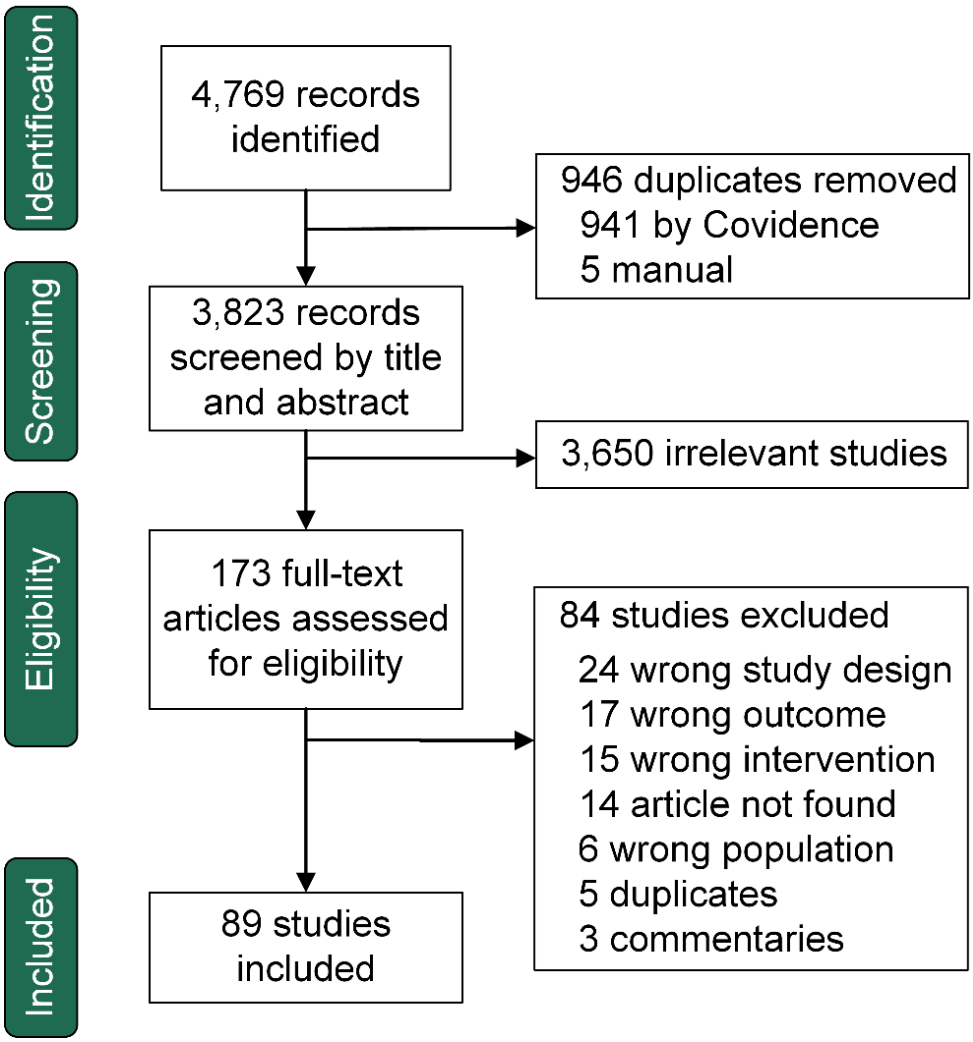

Figure 3. PRISMA flow diagram of study selection.

In brief, from the initial studies, 3,734 articles were found to be irrelevant at either the title and abstract screening or full-text review; data from 89 studies were extracted (Table 1).

Table 1. Description of summarized data from the reviewed articles (n=89).

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
1	Adair, 2022[27]	Home high-intensity interval training	2-10 months of follow up	-	4	3	One group pre + post design	Absolute grip strength, BMI (kg/m^2), and FEV_1%	-	BMI (percentile)
2	Almăjan Guță, 2013[28]	Chest physiotherapy and individualized unsupervised exercise program	12 months	-	4	1	One group pre + post design	BW, skeletal muscle mass, fitness score, power, and force	-	-
3	Andréasson, 1987[29]	Physical exercise training	30 months	-	4	2	One group pre + post design	Productive cough with physical exercise compared with conventional chest PT	Chest X-ray findings, FRC, RV, TLC, and VTG fell in four end-tidal PCO_2 at maximum	FEV_1, Shwachman score VCG was the same in both groups, VC, and ECG
4	Asher, 1982[30]	Inspiratory muscle training	4 weeks of IMT, followed by a 4-week control period	5	2	2	Randomized trial (2 x 2, two-sequence, two-period crossover design)	Inspiratory muscle strength and IM endurance Endurance time	Some patients showed EMG "fatigue" changes in scalene	CO_2, FEF_{25-75}, FEV_1, VC, FRC, or RV/TLC, progressive exercise performance, peak heart rate, peak VO_2, PIM, FRC or PI_{max}
5	Aspinall, 2020[31]	Reduced-exertion high-intensity interval training	6 weeks	-	5	3	Case report	VO_2 peak, GET, OUES, Fatigue index, and QoL scale (Physical, Digestive, Emotional, Treatment, Body Image, and Respiratory domains)		BMI, body mass, and pulmonary function

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
6	Beaudoin, 2017[32]	Intervention: combined aerobic and resistance exercise program.	12 weeks	4	2	2	Open label randomized controlled trial	Glucose Profile, glucose tolerance, insulin sensitivity, Improved PA and maximal strength, and protein levels	-	Inflammatory markers, pulmonary function parameters, and QoL
7	Bieli, 2017[33]	8 weeks of each RME training and standard chest physiotherapy (control period)	8 weeks	6	1	1	Randomized trial (2 x 2, two-sequence, two-period crossover design)	RME test	-	Constant workload cycling test, exercise endurance, FEV_1, FVC, MEF75/25, CF clinical score, and QoL questionnaire
8	Blau, 2002[34]	Intensive 4-week summer camp	4 weeks	-	4	1	One group pre + post design	Dynamic cardiorespiratory parameters during exercise, Peak work capacity, VO_{2max}, VE, VO_2/HR, and the AT	-	Resting vital capacity and maximal expiratory flow rates, cardiorespiratory parameters at rest, the VE/MVV ratio, and the incidence of pan-resistant P aeruginosa or of methicillin-resistant

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
										Staphylococcus aureus
9	Blumquist, 1986[35]	A program of increased physical activity and self-treatment	Study had two parts, each planned for 6 months	-	4	1	One group, 2 phases pre and post design	PA, O_2 pressure, FVC, FEV_1, and small changes in arterialized capillary blood gas analyses in boys		Clinical scores, infection prevalence, X-ray findings, lung function tests, regional lung function, height and weight
10	Braggion, 1989[36]	Aerobic training	8 weeks	-	2	2	Controlled Clinical Trial	Work rate, RPE, R STEADY, the T/2+ of VE, performance, time to exhaustion, and obstacle course completion time	-	-
11	Carr, 2018[37]	Tai Chi lessons	3 months	3	2	2	Randomized controlled feasibility study	General health and wellbeing, children reported trouble sleeping, waking due to cough, daytime cough, trouble breathing, and stomach pain	Questionnaire returns. Adults and teenagers reported worse scores in trouble sleeping, waking due to cough, daytime cough, trouble breathing, the last week.	FVC, FEV_1, oxygen saturation, BMI, The CFQ-R, PSQI, FFMS or CAMM, VAS, general health and wellbeing, the Borg scores, Antibiotic use
12	Cerny, 1989[38]	Exercise therapy + Bronchial hygiene therapy	Duration of hospital stay	4	2	2	Randomized control trial	Peak load, and peak HR, peak HR-to-peak load ratio improved, productive	-	Pulmonary Function Score, sputum volume and dry weight

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
								cough, and pulmonary functions		
13	Clément, 1979[39]	Prone immersion physical exercise: tethered swimming modified by swim fins and a kickboard	28 weeks	-	4	2	One group pre + post design	The FVC, FEV_1, FEV_3, increased in the youngest subject, PWC for exercise capacity, maximal O_2 pulse, respiratory frequency and VO_{2max}. HR_{max} increased in the adolescent girl and young boy. Tidal volume increased for the boys.	The ventilatory equivalent for oxygen increased for the girl and the younger boy.	-
14	de Jong, 1994[40]	Home exercise training program	3 months	-	4	1	One group pre + post design	W_{max}, VO_2max, VO_2/HR VE_{max}, breathing frequency max, HR_{max}, minimal pH values, base excess, HCO_3, anaerobic threshold, the degree of limitation in ADL, and inspiratory vital capacity	-	Pulmonary function, Borg scores, and dead space ventilation
15	de Jong, 2001[41]	Inspiratory muscle training	6 weeks	-	2	2	Controlled Clinical Trial	Inspiratory muscle endurance		FEV_1, FVC, VE_{max}, pI_{max}, w_{max}, $VO_{2\ max}$, Borg & MRC-dyspnea scores, and subjective fatigue scores
16	Dekerlegand, 2014[42]	Home-based inspiratory muscle training program	6 weeks	-	4	3	Case series report	MIP	Peripheral oxygen saturation, dyspnea, and RPE	FEV_1 and 6MWT

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
17	Del Corral, 2017[43]	home exercise program using active video games	6 weeks	8	1	1	Single randomized controlled trial	Walking distance, strength, and HRQoL	-	Annual activity, Horizontal jump, and throw ball
18	de Oliveira, 2009[44]	Aerobic exercise training	6 weeks	5	2	2	Randomized controlled trial	Number of steps, dyspnea, HR, and leg fatigue. C-reactive protein was reduced in G1	-	Resting vital capacity, forced expiratory flow rates, and resting cardiopulmonary parameters
19	de Oliveria, 2017[45]	training program combining aerobic and resistance training	12 weeks	-	4	2	One group pre + post design	FFM, BW, oxygen uptake, and abdominal muscle endurance	-	Fat mass
20	Donadio, 2022[46]	Resistance-training program (EX) alone or EX + neuromuscular electrical stimulation	8 weeks	6	1	1	Randomized controlled trial	VE/VCO_{2peak} and test duration, RER peak, cardiorespiratory fitness TUG, TUDS tests, and peripheral muscle strength (bench press, pectoral, dorsal, and handgrip)	-	QoL
21	Dunlevy, 1994[47]	Structured low impact aerobic exercise program	8 weeks	-	4	2	One group pre + post design	Oxygen consumption, workload, and resting and training heart rate.	-	FEV1, FVC, FEF, and QWB
22	Edlund, 1986[48]	Progressive aerobic swimming program	12 weeks	-	2	1	Controlled Clinical Trial	Shwachman scores, predicted post-VO_{2max}, and exercise time on the treadmill	-	Pulmonary function
23	Elbasan, 2012[49]	Chest physiotherapy and	6 weeks	-	4	1	One group pre + post design	Chest circumference, standing long jump, lateral	-	Sit, reach and FB test

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		aerobic exercise training						flexion, rotation, trunk hyperextension, 20 m shuttle run, 10-step stair climbing, and the parameters related to the treadmill test such as blood pressure, heart rate, inclination and average speed		
24	Elce, 2018[50]	Supervised physical exercise	2 years	-	4	2	Two group pre + post design	Lipid profiles, serum total cholesterol/HDL cholesterol non-HDL, cholesterol/HDL cholesterol ratios, triglycerides, and serum vitamin D levels	Slower decline in Forced FEV_1	SCL, colonization by specific bacteria, CFLD, PI, weight, height, BMI, campesterol, sitosterol, pancreatic enzyme supplementation, serum albumin, vitamin A, vitamin E levels, and the proportion of patients with severe CF (PI)
25	Enright, 2004[51]	High-intensity inspiratory muscle training	8-weeks	5	2	1	Randomized controlled trial	PI_{max}, SPI_{max}, diaphragm thickness during contraction, diaphragm	-	FEV_1, FVC, TFco, inspiratory

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
								thickening ratio, VC, and TLC		muscle function (TDIrel) and diaphragm thickness at FRC and TLC
26	Estévez-González, 2021[52]	Individualized guided resistance	8 weeks	6	1	1	Randomized controlled trial	Muscle strength (bench press, pectoral, and dorsal), cardiorespiratory fitness for HR at 2 minutes of recovery, VO_2 at the AT and VCO_2 at AT, LF power, HF power, and LF/HF ratio) for HRV, time-domain LF/HF ratio, SDNN and RMSSD.	-	
27	Franco, 2014[53]	Pilates mat exercises	16 weeks	-	4	1	One group pre + post design	MIP and MEP (only in females)	-	FVC and FEV_1
28	Goldbart, 2007[54]	Rehabilitation winter camps	3 weeks	-	4	2	One group pre + post design	FVC, FEV_1, Oxyhemoglobin saturation, weight, coughing improved as well as Breathing difficulties	-	-
29	Gruber, 2011[55]	multifaceted rehabilitation program	6 weeks	-	4	1	One group pre + post design	FEV_1, VC, MEF_{25}, MVV, MFL, LFL compared to HFL, fitness level values except for peak fC and at VAT, fitness variables at peak and VAT, and VE/MVV	Lower VE and VT	Better fitness values
30	Gruber, 2014[56]	Intervention 1: interval training treadmill program	6 weeks	-	4	3	Two group pre + post design	Body and fat-free mass, O_{2peak}, $relVO_{2peak}$, VE, and O_2-Pulsepeak,	VC, FEV_1, and P_{peak}	HR_{peak}, VO_2VAT, and HR at VAT

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		intervention2: standard exercise program						PVAT, relPVAT, VEVAT, and O_2-PulseVAT		
31	Gruber, 2008[57]	The supervised training program.	4–6 weeks	-	4	1	One group pre + post design	Weight, BMI, and lung function parameters FEV_1, VC, and MEF_{25}, balancing-bouncing, accurate throw, trunk flexion, standing vertical jumping, and 6MWT	-	For training duration
32	Gruber, 2011[58]	Exercise program	6 weeks	-	4	1	One group pre + post design	Weight, BMI, FEV_1, VC, MEF_{25}, fitness variables, VO_{2peak}, W_{peak}, HR_{peak}, and O_2-Pulsepeak, habitual exercise	-	-
33	Gruber, 2020[59]	Exercise program	12-months	-	4	2	Two group pre + post design	Height, weight, FEV1, FVC, Wpeak, resting HR, higher maximum HR, and DMT subtests except for FB	-	BMI, FEV_1, exercise capacity and HR
34	Gruber, 2021[60]	Physical activities were; sports, traditional sport activities, video games, aerobic exercise, strength training, and games and activities to improve motor/skill performance	2 months	-	4	2	Two group pre + post design	Power or strength, standing long jump, DMT test items based on strength and power, and time spent in activities	PA (steps/day) and PA intensity	Anthropometric data, FEV_1,and W_{peak}

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
35	Gulmans, 1999[61]	Exercise program on a cycle ergometer	6 months	-	4	2	One group pre + post design	Leg muscle strength for knee extensors, and ankle dorsiflexors, VO_{2max}/BM, VO_{2max} /FFM, total Competence Score, as well as the subscales "physical appearance" and "general self-worth".	Perceived acceptability of the program, and willingness to continue cycling	growth, body composition, and energy intake, lung function parameters and Shwachman Scores, strength of the hip extensors, Borg Scale Scores, and willingness to continue with other sorts of training besides cycling
36	Gulmans, 2001[62]	Exercise program on a cycle ergometer	3 months	-	4	1	One group pre + post design	IGF-II, mean IGF-I and the IGF-I/IGFBP-3	-	Clinical and pulmonary condition, fat excretion, and dietary intake, resting energy expenditure, rates of protein degradation, leucine oxidation, and nonoxidative disposal of leucine

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
37	Güngör, 2021[63]	Postural exercise program + chest physiotherapy	6 weeks	7	1	1	Randomized controlled trial	MST distance, respiratory function tests, and the CFQ-R emotional function and treatment difficulties subdomains.	-	All other subdomains of CFQR, limits of stability test items, and cobb and modified Cobb angles.
38	Gupta, 2019[64]	The exercise program of resistance training and plyometric jumping 1 year long	1 year	7	1	1	Single randomized controlled trial	Lumbar spine BMD, maximal oxygen uptake and exercise duration, quality of life-CFQ-R scores, vitamin D levels, number of children colonized with Pseudomona.	Whole body BMD was slightly lower in the experimental Gr compared to controls	FEV_1 %, FVC, habitual activity scores, and QoL
39	Hebestreit, 2010[65]	Sports activities (cycling, strength training, ball games)	1 year	5	2	3	Open label randomized controlled trial	Sports activities, vigorous PA, $VO_{2\ peak}$, time spent in vigorous PA, skinfold thickness, FVC, 'Subjective health perception' on QoL questionnaire.	-	Febrile infections, attitude towards sports activities, PP, MP, height, body mass, FEV_1, and RV/TLC
40	Hebestreit, 2022[66]	Partially supervised conditioning program of strength building exercises aerobic physical activity	1 year	7	1	2	Open label randomized controlled trial	Self-reported vigorous PA, Wpeak, VO_2 peak, greater number of aerobic steps	FEV_1, physician-diagnosed pulmonary exacerbations	The intervention did not impact the time to the first exacerbation or the number of pulmonary exacerbations.

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
41	Heijerman, 1992[67]	Cycle-ergometer	At least 3 weeks	-	4	1	One group pre + post design	FEV, QI, maximal W_{max}, VE_{max}, VO_{2max}, and O_2 pulse	-	VC
42	Heijerman, 1991[68]	Cycle-ergometer	At least 3 weeks	-	4	1	One group pre + post design	FEV, QI, W_{max}, VE_{max}, VO_{2max}, O_2 pulse, and $PaCO_2$ at rest	-	PaO_2 at rest, maximal HR during exercise, and $A\text{-}aDo_2$ at rest and during maximal exercise
43	Holmes, 2022[69]	Home-based resistance training program	12 weeks	-	4	1	One group pre + post design	Reduction in %Fat and FM with a moderate SFBIA displayed opposite trends, while MFBIA showed a similar trend as SFBIA for %Fat and FM, but increase in FFM similar to DXA. An increase in FFM after the intervention.	-	Weight
44	Hommerding, 2015[70]	Aerobic physical exercises + stretching exercises	3 months	5	2	1	Randomized controlled trial	Reported physical exercise practice	-	FVC, FEV_1, FEV_1/FVC, forced expiratory flow of the FVC, body mass, height, BMI, triceps skin fold thickness, arm muscle

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
										circumference, peak VO_2, maximum HR, HR, SpO_2, modified Borg scale, BP, and QoL questionnaire scores: Physical, body image, digestive, respiratory, emotional, social, nutrition, treatment, vitality, health, social role and weight
45	Horvat 1991[71]	Progressive resistance exercise	12 weeks	-	4	2	One group pre + post design	Subject one: physical capacity and self-concept, increased weight, skinfold thickness decreased in ilium, abdomen, and thigh. Girth measurement increased in upper arm, forearm, chest and thigh, hand grip Subject two: Percentages of body fat, self-concept. Increased weight	-	Subject one and two: FVC, RV, TLC, and R pulmonary Subject one alone: percentages of body fat and skinfold thickness

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
								Skinfold thickness decreased in ilium, abdomen, and thigh. Girth measurement increased in the upper arm, forearm, chest and thigh. Hand grip, and abdominal movements increased		
46	Hulzebos, 2011[72]	HIT training program using electronically braked cycle ergometer	6 weeks	-	5	3	Case report	Exercise capacity, FEV_1, RV/TLC, VO_{2peak}, $VO_{2peak/kg}$, W_{peak}, and watt/kg, O_2 pulse, O_2 extraction in muscles, VE_{peak}, breathing depth, respiratory rate, and participation in demanding PA (field hockey)	Ventilatory reserve capacity	-
47	Johnson, 2021[73]	Routine hospital care + encouragment to do aerobic and strength training exercises		-	4	3	Two group pre + post designs	Time spent in exercise and mean estimated exercise per week	-	Estimated minute of exercise, 6MWT distance, and FEV_1
48	Kaltsakas, 2021[74]	Interval or constant-load exercise of electromagnetically braked cycle ergometers	12 weeks	6	1	3	Randomized controlled trial	The IE Gr: quadriceps muscle strength, respiratory muscle strength, and daily PA Both groups: exercise capacity, 6MWT, and QoL	-	-
49	Kaplan, 1992[75]	CF camp (running games, calisthenics, aerobics, water	9 days	-	4	3	One group pre + post design	BW	In year 1 & 2: decrease in PEF and FEF, and 2	PFT parameters

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		sports, field trips, group activities, chest therapy, and meals)							participants harbored pathogenic organisms in sputum	
50	Kenis-Coskun, 2022[76]	Telerehabilitation (based on high-intensity interval training and postural strengthening)	12 weeks	7	1	2	Randomized controlled trial (pilot)	Major depressive disorder subscale, the generalized anxiety disorder score, the body image scores in QoL, and 6MWT	-	QoL scores, caregivers' depression and anxiety scores, the estimated $FEV_{1\%}$ values
51	Klijn, 2004[77]	Anaerobic training	12 weeks	6	1	1	Randomized controlled trial	Absolute PP and MP, in PP and MP per kilogram BW, and in PP and MP per kilogram FFM, Vo_{2peak} W_{max} and serum lactate levels, VO_{2peak} per kilogram, the domain of physical functioning in QoL, mean height, weight, FFM in the TG, and absolute PP	-	Body composition, pulmonary function, peripheral muscle force, and habitual PA, BMI, and the aerobic performance
52	Kriemler, 2013[78]	Strength training + endurance training	6 months	8	1	2	Randomized controlled trial	FVC, FEV_1, RV/TLC, aerobic performance, and VO2 peak	-	Body composition, average-moderate and vigorous PA, muscle power (Wingate test)
53	Ledger, 2013[79]	High intensity interval training using treadmill,	12 months	-	4	2	One group pre + post design	Reduction in treatment and cost in IV, GOSH inpatient IV, shared-care inpatient IV,	Decline in $FEV_{1\%}$	FEV_1 z-scores, growth

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		bike, and cross-trainer + Strength, core-conditioning and stretching components						and home IV, shared-care hospital. Improvement in total CFQ scores, VO_{2Peak}, and MSWT High ratings patients for physiotherapy and dietetic components Improvement in skinfold thickness in some subjects		parameters, BMI z-scores
54	Massery, 2005[80]	The home program interventions were multisystemic (realigning her musculoskeletal system, neuromuscular retraining, joint mobilization, neuromuscular retraining, and strengthening exercises)	4 months	-	5	3	Case report	Postural alignment, thoracic spine movements, chest wall mobility and shape, scapular mobility, resting position Glenohumeral of shoulder, core stabilization of the trunk for postural control and simultaneous support of demanding ventilation maneuvers such as chronic cough, pulmonary function tests, weight, energy, no respiratory exacerbations, and decreased fatigue	-	-
55	McCreery, 2021[81]	Inspiratory Muscle Training using the POWER breathe K5 device	4 weeks	-	4	1	Two groups pre + post design	Decrease in very low frequency post-intervention, and perceived clinically meaningful improvements in respiratory domain	-	HRQoL, Parent/guardian 's perceptions of their child's HRQoL

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
56	McNamara, 2016[82]	One-on-one yoga sessions	10 weeks	-	4	1	One group pre + post design	Anxiety improved in the STAIC, joint pain assessed by APSA, CFQ-R in the domains of emotion and respiratory, self-reported pain evaluated by the MSAS and APSA, and the domain for sleep difficulty by MSAS	-	Sustained anxiety and depression on the CES-DC scale for Children and HADS
57	Moorcroft, 2004[83]	Upper body and lower body exercises	12 months	5	2	2	Randomized controlled trial	Better preservation of pulmonary function, Improvement in FVC, and reductions in ventilation and lactate concentration	-	FEV_1 unchanged
58	O'Neill, 1987[84]	Participants received a copy of exercises graded according to age, sex, and physical fitness, not specific for any form of exercise	2 months	-	4	1	One group pre + post design	RV and breathlessness	Subject 4 indicated a much higher level of breathlessness.	Spirometrics indices, all objective indices of exercise performance at maximal and submaximal workloads, and ventilation. Breathlessness did not change in patient No. 3
59	Orenstein, 1981[85]	Supervised training program (stretching and flexibility	3 months	-	4	2	Two groups pre + post designs	Peak work capacity, peak O_2 consumption, respiratory muscle	-	Pulmonary function

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		exercises + Jog-walk)						endurance, and lower HR for submaximal workloads		
60	Orenstein, 2004[86]	Aerobic Gr + strength training Gr	1 year	7	1	2	Randomized controlled trial	Maximum weight lifted for biceps curls. $VO_{2\ peak}$ in the aerobic Gr after 6 months and Physical W_{peak} FEV_1, 6-12 months Height and weight for both groups.	FEV_1 and VO_{2peak} decreased during the first 6 months	QWB scale
61	Paranjape, 2012[87]	Home exercise regimen consists of regular family-oriented vigorous activities	2 months	-	4	1	One group pre + post design	MSWT, habitual PA, body image perception on QoL scale, and FEV_1. Physical functioning on QoL scale and exercise capacity in boys	-	BMI percentile, FEV_1, and other QoL measures.
62	Prévotat, 2019[88]	The ET program consisted of aerobic training, muscle strengthening, and relaxation.	8 weeks	-	4	1	Two group pre + post designs	FFM, body cell mass, 6MWT distance, bilateral quadriceps muscle strength, exercise tolerance, and muscle strength	-	FEV_1, FVC, BW, BMI, fat mass, brachial circumference, and triceps skinfold.
63	Reuveny, 2020[89]	Ex Gr: HIIT on a cycle ergometer. Control Gr: HIIT while breathing AMB	8 weeks	6	1	1	Randomized controlled trial	Body-fat percentage, decreased BL, "HIIT accelerated low-intensity O_2 uptake kinetics for CWR_{30}". Time to limit tolerance, ventilation, breathing rate decreased, and increased work-interval duration	-	Body mass, lean body mass, pulmonary function measurements, VO_{2peak} or HR_{peak}, training $SpO_{2\%}$

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
64	Rovedder, 2014[90]	Home exercise program based on aerobic training and muscle strength training	3 months	7	1	2	Randomized controlled trial	Muscle strength in the UL,1RM test for left and right UL	-	6MWT, QoL for both the CFQ and SF-36 questionnaires
65	Ruddy, 2015[91]	Standardized yoga program consisting of breathing exercises, postures and relaxation	8 weeks	-	4	1	One group pre + post design	CFQ-R respiratory domain score	-	Other CFQ-R domain scores, CFRSD (respiratory symptom score), FEV_1, weight, and EOBS
66	Sahlberg, 2008[92]	Endurance or resistance training program	6 months	-	2	3	Controlled clinical trial	tcPO2 in males in resistance training Gr. endurance training Gr improved in workload and VO_{2max}	VO_{2max}	RR, HR, VE/VO_2, RER, and FEV_1 after 3 months with ET or RT
67	Sahlberg, 2008[93]	ET was performed using sports (i.e., swimming, jogging, skiing, skating, aerobics or cycling). Patients with RT performed the training at a fitness center. The training followed progressive resistance exercise.	6 months	-	2	3	Controlled clinical trial	$FEV_{1\%}$ in male RT in males for 3 months in right handgrip strength, Serum tocopherol levels increased in ET. The RT Gr increased in IL-6.	In ET, males in muscular strength the ET, decrease in IL-6	FEV_1 over the study period Muscular strength in females Serum tocopherol in RT serum levels of TNFa, IL-1b, and IL-10 in general

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
										$FEV_{1\%}$ or RV in males
68	Santana-Sosa, 2014[94]	Weight-training machines followed by a 4-week detraining period	8 weeks	7	1	1	Randomized controlled trial	PI_{max}, VO_{2peak}, strength tests, body fat percentage, and QoL	-	No desaturation during tests
69	Santana-Sosa, 2012[95]	Weight training machines and cycle ergometers. The second part of the training consisted of strength exercises followed by a 4-week detraining program.	8 weeks	7	1	1	Randomized controlled trial	VO_{2peak}, and strength tests, including 5RM in bench press, leg press, and seated row 5RM	-	FEV_1, FVC, PI_{max}, TUDS, 3-m TUG, weight, BMI, FFM, percentage body fat, and QoL
70	Sartoti, 2008[96]	Respiratory training by SpiroTiger® training device	3 months	-	4	1	One group pre + post design	FEV_1, perception of physical fitness, reduction in the need for IV	-	-
71	Sawyer, 2020[97]	High intensity interval-based cycle ergometry training	8 weeks	7	1	2	Randomized controlled trial	endurance exercise capacity, W_{max}, the physical function domain of the CFQ-R, and the intensity of W_{max}	Four reported muscle soreness	Exercise self-efficacy (BARSE), anxiety and depression (HADS), exercise enjoyment (PACES), & lung function
72	Sawyer, 1993[98]	Inspiratory muscle conditioning trained at pressure	10 weeks	6	1	1	Randomized control trial	PI_{max}, VC, TLC, IC, incremental exercise test.	RV and RV/TLC, hyperinflation, with FRC and TLC	-

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		loads up to 60 % PI_{max}.								
73	Schindel, 2015[99]	Aerobic exercises and stretching guidelines	3 months	6	1	1	Randomized control trial	Cervical & lumbar lordosis, thoracic kyphosis, lateral chest distance, abdominal protrusion, and distribution of plantar pressures	-	-
74	Schmidt, 2011[100]	Individually tailored exercise program where subjects chose bicycling, running, gymnastics, swimming, and dancing	12 weeks	-	4	3	One group pre + post design	VO_{2max}, domain of treatment burden and emotional functioning in QoL	-	Total QoL score, lung function $FEV_{1\%}$, and BMI
75	Schneiderman-Walker, 2000[101]	Aerobic activities	3 years	6	1	1	Randomized control trial	Engagement in PA, feeling better about themselves, sense of well-being, more energy, and less chest congestion	FVC & FEV_1	Exercise parameter, x-ray or Shwachman scores, hospitalizations
76	Selvadurai, 2002[102]	Aerobic activities + resistance training	Duration of hospital stay	5	2	1	Randomized control trial	AT: peak aerobic capacity, activity levels, and QoL RT: weight, total mass, FFM, lung function, and leg strength	RT: experience muscle strength loss after discharge	In both groups: FVC & O_2 desaturation
77	Shaw, 2016[103]	Progressive RT program	4 weeks	-	4	1	Two groups + post design	FEV_1, FEV_1/FVC, latissimus dorsi, pectoralis strength, emotional and QoL domains on the CFQ-R	Pectoralis minor strength on the left, and social, body image, and respiration, QoL	-

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
									domains on the CFQ-R in the RTG	
78	Strauss, 1987[104]	Variable weight training	6 months	-	4	2	One group pre + post design	RV, the RV/TLC ratio, chest diameter changed both AP and transversely, BW, muscle size, strength in the upper arms, pulmonary function, and self-reported positive experience	-	-
79	Tomlinson, 2020[105]	Video calls (supervised exercises: bikes, treadmills, with others using free-weights, resistance-bands or bodyweight exercises)	8 weeks	-	4	1	One group pre + post design	Positive feedback on Skype for exercise and high satisfaction with the research study	-	Body size, lung function, PA, or QoL variables No adverse events
80	Turchetta, 2004[106]	Training program consisted of walking, running on the treadmill	12 weeks	-	4	2	Oe group pre + post design	TE, VO_2, $VO_{2/kg}$, and pulmonary ventilation. Reductions in cost and hospital IV days and home IV	-	FEV_1, FVC, HR_{max}, and SBPm
81	Urquhart, 2012[106]	Home exercise and airway clearance program including aerobic exercise, strength training,	1 year	-	4	1	One group pre + post design	cost reduction of £66,384. Increase in MST, absolute FEV_1, QOL: physical, emotional, social, body, treatment, and respiratory domains	-	$FEV_{1\%}$ predicted and lung function

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
82	Van Biervliet, 2021[107]	Physical activity: swimming and fitness training	3 weeks	-	4	2	One group pre + post design	BW, BMI z-score, FFMI, FMI, $FEV_{1\%}$ and $FVC_{\%}$ Number of malnourished patients reduced	-	-
83	van de Weert-van Leeuwen, 2014[108]	Home-based exercise program consisting of callisthenic exercises (stretching, sit-ups, back extension, push-ups) and the fifth is an aerobic exercise (running)	12 weeks	-	4	3	Two groups pre + post design	Better training response, and low inflammation infection status Exercise capacity for those with lower inflammation Lung function improvement in patients with P. aeruginosa colonization	-	Aeruginosa infe ction status, aeruginosa colonization, and BMI
84	Vivodtzev, 2013[109]	Neuromuscular Electrical Stimulation training program followed by ergo cycle (ERGO) training on a cycle ergometer	8 weeks	4	2	3	Randomized control trial	Work rate, BMI, maximal HR, maximal aerobic capacity, mid-thigh circumference and quadriceps strength, fasting glucose and C peptide, procalcitonin, HOMA-R, physical function and health perception CFQ1 scores, and the Transition Dyspnea Index	-	6MWD, lung function, other inflammatory parameters, leg fatigue or dyspnea scores, and blood composition except for procalcitonin, and the HOMA-R of pancreatic b - cell function
85	Welsner, 2021[110]	Individual exercise program: aerobic	12 months	-	4	1	One group pre + post	Health-related and motor performance fitness test scores	-	W_{peak}, HPA, BMI, and $ppFEV_1$

	Primary author, Year	Intervention type	Duration	Quality scores			Study type	Outcome measures		
				PEDro scale	Modified Sackett's levels of evidence	EPHPP quality assessment tool		Improvement	Decline	No change
		training + resistance training								
86	YalÇInkaya, 2019[111]	Pulmonary rehabilitation program: aerobic training+ stretching + breathing exercises.	8 weeks	-	5	3	Case report	The sputum, severity of dyspnea perception (MMRC: 0) and cough symptoms, 6-MWT, FEV_1, FVC, FEV_1/FVC, workload, VO_{2max}, SpO_2, HR, the grip strength, static trunk endurance, and HRQoL parameters	-	Quadriceps muscle strength
87	Zach, 1981[112]	Swimming lessons	7.5 weeks	-	4	2	One group pre + post design	FVC, FEV_1, sputum volume, FEF_{25-75}, and PEFR	-	FEV_1/FVC
88	Zach, 1982[113]	Vigorous activities: swimming, jogging, gymnastics, skipping, and a choice to play minigolf, soccer, and/or table tennis	17 days	-	4	1	One group pre + post design	FVC, FEV, FEF, PEFR, coughing and volume of expectorated sputum	-	FEV_1, FEF_{25-75}, TLC, RV, and RV/TLC
89	Zeren, 2019[114]	Chest PT Gr + moderate-intensity physical activity: brisk walking, sports, dancing, ball games, simple exercises on Pilates ball + Inspiratory Muscle Training	8 weeks	7	1	2	Randomized controlled trial	Overall score of LOST PT+IMT and PT, FEV_1, PEF, MIP, and MEP, 6MWT and improved O_2 desaturation, and engagement in PA	-	PST and directional scores of LOST

Table legend: PEDro, Physiotherapy Evidence Database; EPHPP, The Effective Public Health Practice Project; BMI, body mass index; FEV_1%, forced expiratory volume in one second; (kg/m^2), kg, kilograms ; m^2 meters squared; RV, residual volume; TLC, total lung capacity; VTG, vital to tidal gas ratio; FVC, forced vital capacity; VC, vital capacity; CF, cystic fibrosis; AT, anaerobic threshold; VAS, visual analog scale; RME, respiratory muscle endurance; RPE, rate of perceived exertion; ET, endurance training; RT, resistance training; PT, physiotherapy; PA, physical activity; FFM, fat-free mass; PEF/PEFR, peak expiratory flow (R) rate; 6-MWT/D, 6-minute walk test /distance; BP, blood pressure; ERGO, cycle ergometer; AMB, breathing ambient air; EX, exercise; BW, body weight; PCO_2, partial pressure of carbon dioxide; VCG, vectorcardiography; ECG, electrocardiogram; IM, inspiratory muscle; IMT, inspiratory muscle training; EMG, electromyography; CO_2, carbon dioxide; FEF, forced expiratory flow; VO_2,oxygen consumption; PI_{max}, maximal inspiratory pressure; OUES, oxygen uptake efficiency slope; VO_2, peak volume of oxygen uptake during peak exercise; GET, gas exchange threshold; QoL, quality of life; MEF75/25, maximal expiratory flows at 75/25% of vital capacity; VO_2/heart rate, oxygen pulse; V_E/MVV ratio, dyspnea index; R STEADY, respiratory exchange ratio steady state; T/2+ of V_E, the half time of the peak minute ventilation; CFQ-R, the Cystic Fibrosis Questionnaire-Revise; PSQI, the Pittsburgh Sleep Quality Index; FFMS, the Five Facet Mindfulness Scale; CAMM, the Child and Adolescent Mindfulness Measure; FEV_3, forced expiratory volume in three seconds; PWC, physical work capacity;; Max, maximum; V_{Emax}, maximal minute ventilation; HCO_3, bicarbonate; ADL, activities of daily living; Borg, the Borg rating of perceived exertion; MRC, Medical Research Council (dyspnea scale); HRQoL health-related quality of life; G1, group 1; V_E/VCO2, minute ventilation to carbon dioxide production ratio; RER, respiratory exchange ratio; TUG, timed up and go test; TUDS, tests timed up and down stairs test; 20 m, 20-meter; SCL, Sweat chloride levels; CFLD, cystic fibrosis liver disease; PI, pancreatic insufficiency; HDL, high-density lipoprotein; SPI_{max}, sustained maximal inspiratory pressure; TFco, transfer factor of the lung for carbon monoxide; TDIrel, diaphragm thickness measured by ultrasound at functional residual capacity; VCO_2, volume of carbon dioxide output; LF, low frequency; HF, high frequency; LF/HF ratio, low-frequency to high-frequency ratio; HRV, heart rate variability; SDNN, standard deviation of R-R intervals (R waves); RMSSD, root mean square of successive differences between normal heartbeats; MVV, maximum voluntary ventilation; MFL, middle fitness level group; LFL, low fitness level group; HFL, high fitness level group; fC, cardiac frequency; VAT, ventilatory anaerobic threshold; VT, tidal volume; HR_{peak}, peak heart rate; VO_2VAT, oxygen uptake at the ventilatory anaerobic threshold; P_{peak}, peak power; O_{2peak}, peak oxygen uptake; relVO_{2peak}, the peak oxygen uptake related to body weight; V_E, minute ventilation; O_2-pulsepeak, peak oxygen pulse; PVAT, peak ventilatory anaerobic threshold; relPVAT, peak ventilatory anaerobic threshold to body weight; V_EVAT, ventilatory equivalent for oxygen at the anaerobic threshold; O_2-pulseVAT, oxygen pulse at the ventilatory anaerobic threshold; FB, forward bend; DMT, test Deutscher Motorik Test; W_{peak}, peak workload/work capacity; IGF-II, insulin-like growth factor II; IGF-I, Insulin-like growth factor I; IGF BP-3, insulin-like growth factor binding protein 3; MST/ MSWT, modified shuttle test; BMD, bone mineral density; RV/TLC, residual volume relative to total lung capacity; VO_{2max} ,maximal oxygen uptake; QI, Quetelet Index; Wmax, maximal work capacity; $PaCO_2$, partial pressure of carbon dioxide in arterial blood; PaO_2,partial pressure of oxygen in arterial blood; A-aDo2, alveolar-arterial difference in peripheral capillary oxygen; FM, fat mass; SFBIA, segmental bioelectrical impedance analysis; MFBIA, multi-frequency bioelectrical impedance analysis; DXA, dual energy X-ray absorptiometry; FEV_1/FVC, forced expiratory volume in one second/forced vital capacity; HR_{max}, maximum heart rate; HR, heart rate; SpO_2, peripheral capillary oxygen saturation; R pulmonary, resistance pulmonary;; watt/kg, V_{Epeak}, peak ventilation; IE Gr, interval exercise group; PFT, pulmonary function test; MP, mean power; PP, peak power; TG, training group; IV, intravenous; GOSH, Great Ormond Street Hospital for Children; CFQ, Cystic Fibrosis Questionnaire; STAIC, the Spielberger State-Trait Anxiety Inventory for Children; MSAS, the Memorial Symptom Assessment Scale; APSA, the Additional Pain Symptoms Assessment; CES-DC, Center for Epidemiological Studies-Depression Scale for Children; HADS, Hospital Anxiety and Depression Scale; QWB, Quality of Well-Being scale; ET, exercise training; Ex Gr, exercise group; BL, blood lactate; CWR, constant work rate; SF-36 , 36-Item Short Form urvey; UL, upper limb; CFRSD, Cystic Fibrosis Respiratory Symptom Diary; EOBS, ease of breathing score; HIT/HIIT, high-intensity interval training; RR, respiratory rate; V_E/Vo_2, ventilatory equivalent for oxygen; $tcPO_2$, transcutaneous oxygen tension; IL-6, interleukin-6; TNFa, tumor necrosis factor-alpha; IL-1b, interleukin-1 beta; IL-10, interleukin-10; TUDS, Timed Up and Down Stairs; TUG, Timed Up-and-Go; 5RM, 5 repetition maximum; BARSE, barriers self-efficacy scale; PACES, Physical Activity Enjoyment Scale; FRC, functional residual capacity; IC, inspiratory capacity; RT, resistance training; RTG, resistance training group; AP, anterior posterior; SBPm, maximal systolic blood pressure; TE, time of exercise; FFMI, fat-free mass index; FMI, fat mass index; C peptide, connecting peptide; HOMA-R, the homeostasis model assessment index of insulin resistance; b-cell, beta cell; HPA, habitual physical activity; $ppFEV_1$, percent predicted FEV_1; MMRC: Modified Medical Research Council; PST, Postural Stability Test; LOST, Limits of Stability Test; MIP, maximum inspiratory pressure; MEP, maximum expiratory pressure.

Of the eligible studies, 36% were randomized-controlled trials, 6% were controlled clinical trials, 11% were two group pre-post designs, 41% were one-group pre-post designs, and 6% were either case series or case reports. The most used mode of exercise was aerobic activity followed by resistance training and respiratory exercise. The duration of study interventions ranged between one week to 3 years. The sessions ranged from 2-7 per week, generally lasting from 45 minutes to an hour. In general, the improvement after the exercise interventions were noted mostly in exercise capacity, pulmonary function, quality of life, muscle strength, and body composition.

3.1. OUTCOME MEASURES AND THE INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY AND HEALTH (ICF) CODES

The total number of extracted outcomes was 1,116, with many of these extracted from survey/questionnaire items. Tables 1-4 illustrate the ICF categories that were targeted by the selected studies. In these tables, the frequency represents the number of codes addressed by the studies. Each code was addressed once per study, regardless of the frequent link to multiple outcome measures in the same study. Overall, 1,047 outcome measures were linked to 108 ICF categories at the first and second levels. Refer to Figure 4 for the distribution of outcome measures based on ICF domains. There were 69 outcome measures that were either "nd" (not definable) or "nc" (not covered). These outcome measures are referred to as "Others" in Figure 4.

The questionnaire items that were used frequently by the studies in this review are presented in an appendix table (Appendix). The scales in this table included the Cystic Fibrosis Questionnaire/Cystic Fibrosis Questionnaire-Revised, Fatigue Index, Physical Activity Recall, Hospital Anxiety and Depression Scale, and the Quality of Well-Being Scale.

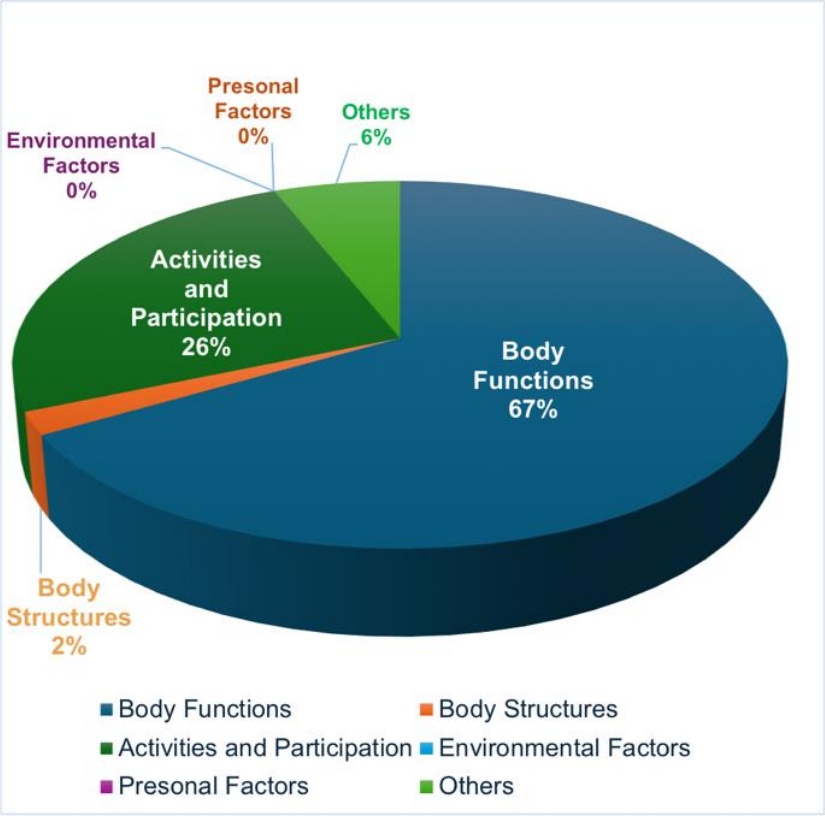

Figure 4. Percentage of ICF categories linked to the outcome measures of the studies.

Seven of eight "chapters," or domains, at the Body Functions level were addressed (Table 2). A wide variety of categories within each domain were assessed. The only domain not covered by any outcome measure was "Chapter 3: Voice and speech functions."

Table 2. The frequency of occurrences in the coded outcome measures (the first and second-level ICF categories) for Body Functions.

ICF code	Description	Frequency
Body Functions		
Chapter 1: Mental functions		
b110	Consciousness functions	1
b114	Orientation functions	1
b126	Temperament and personality function	16
b130	Energy and drive functions	53
b134	Sleep functions	6
b140	Attention functions	3
b152	Emotional functions	31
b156	Perceptual functions	1
b167	Mental functions of Language	1
b180	Experience of self and time functions	33
Chapter 2: Sensory functions and pain		
b235	Vestibular functions	6
b240	Sensation associated with hearing and vestibular function	1
b250	Taste functions	1
b280	Sensation of pain	13
Chapter 4: Functions of the cardiovascular, hematological, immunological and respiratory systems		
b410	Heart function	35
b420	Blood pressure functions	9
b430	Hematological system functions	24
b440	Respiratory function	87
b445	Respiratory muscle functions	11
b450	Additional respiratory functions	16
b455	Exercise tolerance functions	138
b460	Sensations associated with cardiovascular and respiratory functions	38
Chapter 5: Functions of the digestive, metabolic and endocrine systems		
b510	Ingestion functions	2
b515	Digestive functions	26
b525	Defecation functions	26
b530	Weight maintenance functions	62
b535	Sensations associated with the digestive system	25
b540	General metabolic functions	3
b545	Water, mineral and electrolyte balance functions	4
b550	Thermoregulatory functions	3
b599	Functions of the digestive, metabolic and endocrine systems, unspecified	2
Chapter 6: Genitourinary and reproductive functions		
b620	Urination functions	1
b640	Sexual functions	1
Chapter 7: Neuromusculoskeletal and movement-related functions		
b710	Mobility of joint functions	3
b720	Mobility of bone functions	2
b730	Muscle power functions	34
b740	muscle endurance function	11
b770	Gait pattern functions	1
Chapter 8: Functions of the skin & related structures		
b810	Protective functions of the skin	1
b830	Other functions of the skin	1
b840	Sensation related to the skin	1

For Body Structures, only two of the eight domains were assessed by outcome measures (Table 3).

Table 3. The frequency of occurrences in the coded outcome measures (the first and second-level ICF categories) for Body Structures.

ICF code	Description	Frequency
Body Structures		
Chapter 4: Structures of the cardiovascular, immunological and respiratory systems		
s430	Structure of respiratory system	6
Chapter 7: Structures related to movement		
s710	Structure of head and neck region	1
s730	Structure of upper extremity	5
s740	Structure of pelvic region	1
s750	Structure of lower extremity	4
s760	Structure of trunk	9

At the Activities and Participation level, seven of nine chapters (domains) were addressed by various categories of outcome measures (Table 4). The only domains not covered were Learning and applying knowledge, and Communication.

Table 4. The frequency of occurrences in the coded outcome measures (the first and second-level ICF categories) for Activities and Participation.

ICF code	Description	Frequency
Activities and Participation		
Chapter 2: General tasks and demands		
d230	Carrying out daily routine	3
Chapter 4: Mobility		
d410	Changing basic body position	9
d430	Lifting and carrying objects	2
d445	Hand and arm use	2
d450	Walking	13
d455	Moving around	6
d460	Moving around in different locations	1
d465	Moving around using equipment	1
d470	Using transportation	3
Chapter 5: Self-Care		
d510	Washing oneself	1
d540	Dressing	3
d570	Looking after one's health	69
Chapter 6: Domestic Life		
d620	Acquisition of goods and services	19
d640	Doing housework	2
Chapter 7: Interpersonal and interactions		
d7	Interpersonal interactions and relationships	25
d710	Basic interpersonal interactions	1
d740	Formal relationships	1
d750	Informal social relationships	2

ICF code	Description	Frequency
b770	Intimate relationships	1
Chapter 8: Major life areas		
d8	Major life areas	24
d820	School education	20
d845	Acquiring, keeping and terminating a job	19
d850	Remunerative employment	2
d855	Non-remunerative employment	1
d860	Basic economic transactions	19
Chapter 9: Community, Social and Civic life		
d9	Community, social and civic life	24
d920	Recreation and leisure	10

For Environmental Factors, only one out of five domains was addressed by only two studies (Table 5).

Table 5. The frequency of occurrences in the coded outcome measures (the first and second-level ICF categories) for Environmental Factors.

ICF code	Description	Frequency
Environmental Factors		
Chapter 4: Attitudes		
e410	Individual attitudes of immediate family members	1
e460	Societal attitudes	1

3.2. METHODOLOGICAL QUALITY

The PEDro score results for the 32 studies ranged from 3 to 8, with a median of 6. For the modified Sackett's level of evidence, only 21 of 89 studies were ranked as level 1. Using the Quality Assessment Tool for Quantitative Studies (the EPHPP) 49% were ranked as high quality, 34% moderate, and 17% were low, with a median of 2 (Moderate) (Figure 5 and Table 6).

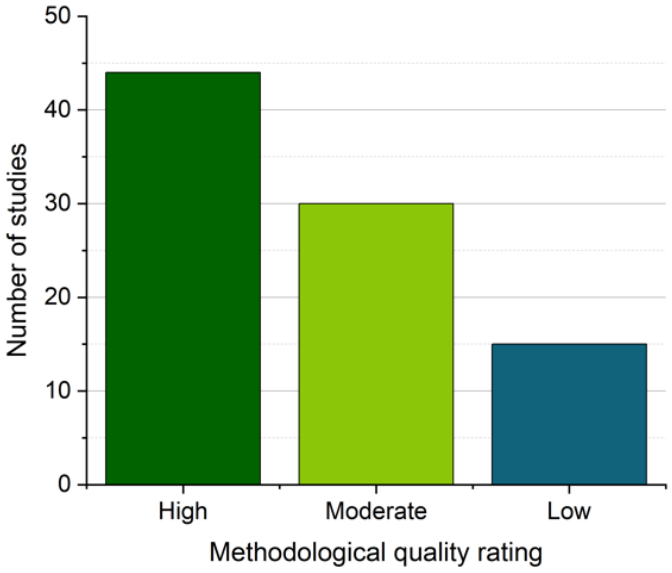

Figure 5. Methodological quality rating distribution using the EPHPP scale for quantitative studies.

Table 6. The effective public health practice project (EPHPP) quality assessment tool scoring results.

	First author, Year	Selection bias	Study design	Confounders	Blinding	Data collection methods	Withdrawal and dropout		Overall rating*
1	Adair, 2022	Weak	Moderate	Strong	Moderate	Strong	Weak		Weak
2	Almăjan Guță, 2013	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
3	Andréasson 1987	Moderate	Moderate	Weak	Moderate	Strong	Strong		Moderate
4	Asher, 1982	Moderate	Strong	Weak	Moderate	Strong	Moderate		Moderate
5	Aspinall, 2020	Weak	Weak	Strong	Moderate	Strong	Moderate		Weak
6	Beaudoin, 2017	Strong	Strong	Strong	Weak	Strong	Moderate		Moderate
7	Bieli, 2017	Moderate	Strong	Strong	Moderate	Strong	Moderate		Strong
8	Blau, 2002	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
9	Blomquis, 1986	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
10	Braggion, 1989	Moderate	Strong	Weak	Moderate	Strong	Strong		Moderate
11	Carr, 2018	Weak	Strong	Strong	Moderate	Strong	Moderate		Moderate
12	Cerny, 1989	Moderate	Strong	Weak	Moderate	Strong	Strong		Moderate
13	Clement, 1979	Moderate	Moderate	Strong	Moderate	Strong	Weak		Moderate
14	DeJong, 1994	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
15	DeJong, 2001	Moderate	Strong	Weak	Moderate	Strong	Strong		Moderate
16	Dekerlegand, 2014	Weak	Weak	Strong	Moderate	Strong	Strong		Weak
17	DelCorral, 2017	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
18	DeOliveira, 2009	Moderate	Strong	Strong	Moderate	Strong	Moderate		Strong
19	DeOliveira, 2017	Weak	Moderate	Strong	Moderate	Strong	Strong		Moderate
20	Donadio, 2022	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
21	Dunlevy, 1994	Weak	Moderate	Strong	Moderate	Strong	Moderate		Moderate
22	Edlund, 1986	Moderate	Strong	Strong	Moderate	Moderate	Strong		Strong
23	Elbasan, 2012	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
24	Elce, 2018	Moderate	Moderate	Strong	Moderate	Strong	Weak		Moderate
25	Enright, 2004	Moderate	Strong	Strong	Strong	Strong	Strong		Strong
26	Estevez-Gonzalez, 2021	Moderate	Strong	Strong	Moderate	Strong	Moderate		Strong
27	Franco, 2014	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
28	Goldbart, 2007	Moderate	Moderate	Strong	Moderate	Strong	Weak		Moderate
29	Gruber, 2011	Moderate	Moderate	strong	Moderate	Strong	Moderate		Strong
30	Gruber, 2014	Weak	Moderate	Weak	Moderate	Strong	Strong		Weak
31	Gruber, 2008	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
32	Gruber, 2011	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
33	Gruber, 2020	Moderate	Moderate	Weak	Moderate	Strong	Moderate		Moderate

34	Gruber, 2021	Moderate	Moderate	Weak	Moderate	Strong	Moderate		Moderate
35	Gulmans, 1999	Weak	Moderate	Strong	Moderate	Strong	Strong		Moderate
36	Gulmans, 2001	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
37	Gungor, 2021	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
38	Gupta, 2019	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
39	Hebestreit, 2010	Moderate	Strong	Weak	Weak	Strong	Moderate		Weak
40	Hebestreit, 2022	Strong	Strong	Strong	Weak	Strong	Strong		Moderate
41	Heijerman, 1992	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
42	Heijerman, 1991	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
43	Holmes, 2022	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
44	Hommerding, 2015	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
45	Horvat & Carlile, 1991	Weak	Moderate	Strong	Moderate	Strong	Strong		Moderate
46	Hulzebos, 2011	Weak	Weak	Strong	Moderate	Strong	Moderate		Weak
47	Johnson, 2021	Moderate	Moderate	Weak	Moderate	Strong	Weak		Weak
48	Kaltsakas, 2021	Weak	Strong	Weak	Moderate	Strong	Strong		Weak
49	Kaplan, 1992	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
50	Kenis-Coskun, 2022	Weak	Strong	Strong	Moderate	Strong	Strong		Moderate
51	Klijn, 2004	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
52	Kriemler, 2013	Moderate	Strong	Weak	Moderate	Strong	Moderate		Moderate
53	Ledger, 2013	Moderate	Moderate	Weak	Moderate	Strong	Moderate		Moderate
54	Massery, 2005	Weak	Weak	Strong	Moderate	Strong	Moderate		Weak
55	McCreery, 2021	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
56	McNamara, 2016	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
57	Moorcroft, 2004	Weak	Strong	Strong	Moderate	Strong	Strong		Moderate
58	O'Neill, 1987	Moderate	Moderate	Strong	Strong	Strong	Strong		Strong
59	Orenstein, 1981	Weak	Moderate	Strong	Moderate	Strong	Strong		Moderate
60	Orenstein, 2004	Moderate	Strong	Strong	Moderate	Strong	Weak		Moderate
61	Paranjape, 2012	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
62	Prevotat, 2019	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
63	Reuveny, 2020	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
64	Rovedder, 2014	Moderate	Strong	Weak	Moderate	Strong	Strong		Moderate
65	Ruddy, 2015	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
66	Sahlberg, 2008	Weak	Strong	Weak	Moderate	Strong	Strong		Weak
67	Sahlberg. 2008	Weak	Strong	Weak	Moderate	Strong	Strong		Weak
68	Santana-Sosa, 2014	Moderate	Strong	Strong	Strong	Strong	Strong		Strong
69	Santana-Sosa, 2012	Strong	Strong	Strong	Strong	Strong	Strong		Strong
70	Sartoti, 2008	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong

71	Sawyer, 2020	Weak	Strong	Strong	Strong	Strong	Strong		Moderate
72	Sawyer, 1993	Moderate	Strong	Strong	Strong	Strong	Strong		Strong
73	Schindel, 2015	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
74	Schmidt, 2011	Weak	Moderate	Strong	Weak	Strong	Weak		Weak
75	Schneiderman-Walker, 2000	Moderate	Strong	Strong	Strong	Strong	Strong		Strong
76	Selvadurai, 2002	Moderate	Strong	Strong	Moderate	Strong	Strong		Strong
77	Shaw, 2016	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
78	Strauss, 1987	Weak	Moderate	Strong	Moderate	Strong	Moderate		Moderate
79	Tomlinson, 2020	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
80	Turchetta, 2004	Weak	Moderate	Strong	Moderate	Strong	Strong		Moderate
81	Urquhart, 2012	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
82	Van Biervliet 2020	Weak	Moderate	Strong	Moderate	Strong	Strong		Moderate
83	van de Weert-van Leeuwen, 2014	Weak	Moderate	Weak	Moderate	Strong	Moderate		Weak
84	Vivodtzev, 2013	Weak	Strong	Weak	Moderate	Strong	Strong		Weak
85	Welsner, 2021	Moderate	Moderate	Strong	Moderate	Strong	Moderate		Strong
86	YalÇInkaya, 2019	Weak	Weak	Strong	Moderate	Strong	Moderate		Weak
87	Zach, 1981	Moderate	Moderate	Strong	Moderate	Strong	Weak		Moderate
88	Zach, 1982	Moderate	Moderate	Strong	Moderate	Strong	Strong		Strong
89	Zeren, 2019	Weak	Strong	Strong	Strong	Strong	Strong		Moderate

4. Discussion

This review was the first to use the ICF model in relation to exercise intervention studies in people with CF. In general, the improvements across all the studies' outcomes outweighed the declines, mostly in exercise capacity, quality of life, muscle strength, pulmonary function, body composition, and relief of symptoms. These improvements are similar to those reported in a Cochrane review by Radtke et al in 2022[7]. However, it is worth noting that in advanced conditions, some outcome measures declined in number of patients, which underlines the significance of implementing individualized intervention plans to enhance the sustainability of health improvements. Such an individualized approach is also encouraged by other authors[115,116].

Based on the findings of this review, the outcome measures of the included studies mostly targeted the domain of Body Functions, see figure 5. These results are supported by existing research in literature. According to a Cochrane review in CF, most of the exercise outcomes of the studies in their review were addressing the domain of Body Functions.[7] However, this review did not utilize the ICF model. Fortunately, we found other reviews that utilized the ICF model in relation to exercise, however, these reviews were conducted in people with different health conditions. Based on these reviews, outcome measures of exercise studies were mostly related to the domain of Body Functions. The findings Silverman et al[9] and Lima et al[117] also support our findings.

As anticipated, the most frequent code within the Body Functions in this review was respiratory functions (b440), followed by exercise tolerance functions (b455), and weight maintenance functions (b530). These codes covered several outcome measures such as forced expiratory volume in 1 second (FEV_1), forced vital capacity (FVC), peak workload (W_{peak}), maximum oxygen uptake (VO_{2max}), body mass index (BMI), and skinfold thickness. There were also other frequently addressed outcomes within this domain which included: energy and drive functions (b130), sensations associated with cardiovascular and respiratory functions (b460), heart function (b410), and muscle power functions (b730). The codes covered heart rate, rate of perceived exertion, 5-repetition maximum test, handgrip strength, jump tests, and questionnaire items such as "You felt full of energy." Since the most affected system in CF is the pulmonary system, these results were compared to the core sets for chronic obstructive pulmonary disease (COPD)[118]. In line with the findings in this study, respiratory functions (b440), exercise tolerance functions (b455), and muscle power functions (b730) were frequently addressed in people with COPD[119].

The Activities and Participation domain was commonly coded within the studies' outcome measures. The most frequent occurrence was looking after one's health (d570), which also includes managing diet and fitness (d5701). This is due to the heavy use of CF health-related quality of life and physical activity questionnaires by the studies. Both codes cover the following items: performing physical activities, time spent each day on treatments, and difficulty doing treatments (including medications) each day. The most frequent code within the Body Structure domain was structure of trunk (s760). This code covered bone mineral density for lumber spine (BMD) as well as chest and trunk postural evaluation. The Environmental Factors domain included individual attitudes of immediate family members (e410) and societal attitudes (e460). They covered two questionnaire items, which were "People were unfriendly" and "My family is disappointed in me." However, the two ICF categories were addressed only by two studies. The impact of exercise interventions on body functions can be affected by environmental factors such as climate and the availability of exercise equipment. However, these outcome measures were not assessed by the studies in this review. This statement is supported by Mandrusiak's study, which emphasizes the effect of contextual factors (e.g., environmental) on functioning in people with CF[120].

The Personal Factors "pf" in this review included self-efficacy, self-perception, and preference of

exercise. The coding of these personal factors was based on the ICF core set for obstructive pulmonary diseases[118]. The coding of "nd" and "nc" included hospitalization, inflammatory markers, antibiotic use, sputum culture, and general mental and physical health. For surveys, the most frequently used questionnaires were the "CF Questionnaire" (CFQ) and "CFQ-Revised" (CFQ-R). The updated rules of the linking process were used to code the questionnaire items in the studies included. Based on the updated rules, the main and additional concepts should be linked to the ICF codes.[17] For example, in the item "To what extent do your treatments make your daily life more difficult," the main concept was 'treatments' and was linked to looking after one's health (d570) and the additional concept was 'daily life' which was linked to carrying out daily routine (d230) (Appendix). The main concept from all questionnaire items were also addressed in Tables 2-5.

Although the outcome measures of the selected studies covered many codes, most of these studies targeted similar outcome measures. The study of Mandrusiak also supports this information. According to this study, the most frequently targeted functions in people with CF are related to the pulmonary and digestive systems[120]. One of the least addressed outcome measures in this review was urinary incontinence, even though this is a not an uncommon issue in people with CF[121,122], which can be managed by specialized exercise training with a physical therapist[123].

According to the EPHPP, the median analysis for the study's quality was 2 which is a moderate rating. The results from the modified Sackett's level of evidence also support the EPHPP findings. Only 21 studies were qualified as high level of evidence. The frequent rating of weak for included studies based on the EPHPP was due to selection bias, confounders, and withdrawals, respectively, see Table 6.

4.1. STRENGTHS AND LIMITATIONS

It is worth mentioning that this review had a number of strengths, as it can provide a standardized framework assessment for CF and guide comprehensive patient care. There are also several limitations to this review. According to the EPHPP scale, some of the six main items were not applicable to one group pre-post studies. In the confounder section, for instance, the question of whether there are important differences between groups doesn't apply to one group pre + post designs. To avoid rating the section as weak in this case, we selected "no", which might have biased the rating. Also, since the inclusion criteria were not limited to a specific period, several included studies were conducted over 40 years, and we were unable to find 14 articles. This is considered a limitation in this review since the ICF was first introduced in 2001. Lastly, several questionnaires used in studies could not be found, thus, we were unable to link all questionnaire items to ICF codes.

5. Conclusion

There are many proponents that "Exercise is Medicine in Cystic Fibrosis"[124]. Our study, one of the largest to evaluate exercise outcomes in cystic fibrosis regardless of study type would also support that in CF, Exercise is Medicine®[125]. Various types of exercise was found to improve aerobic exercise tolerance, strength, posture, body composition, as well as quality of life. However, exercise studies for individuals with CF have mostly targeted the domains of "Body Functions" and "Activities and Participation." Also, the methodological quality of exercise studies included in this review are generally moderate.

6. Future directions

This review informs future exercise studies in CF studies to target more categories within the ICF framework, such as the structure of the cardiovascular system, to reflect the potential effects of exercise on body structure in people with CF. It is also important to have people with CF engaged in the study design to ensure that parameters that are important to them are being assessed in clinical trials. Based on the results of this review, researchers should adhere to more

vigorous methodological designs to minimize the risk of bias and improve the quality of exercise studies in CF. Lastly, it was often unclear exactly what the intervention consisted of or how it was implemented and progressed; hence, we suggest use of the Template for Intervention Description and Replication (TIDieR) checklist and guide for future investigators so that their intervention can be replicated, especially with the new era of highly effective modulator therapies, in which more people with CF should be able to exercise[126].

7. Conflict of Interest:

The authors have no conflicts of interest to declare.

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Appendix.

Table of most frequently used questionnaires, illustrating questionnaire items and linking to ICF categories.

1. Name of instrument or other identifier	2. Verbatim health information (e.g., wording of item or instruction)	3. Perspective adopted in information	4. Response options	5. Classification of response options	6. Main concept: What is this information about?	7. Additional concepts contained in information	8. ICF category of main concept	9. ICF category of other concepts	10. Annotation
Cystic Fibrosis Questionnaire/Cystic Fibrosis Questionnaire Revised[127,128]									
Item 1 of physical functioning	Performing vigorous activities such as running or playing sports	Descriptive: Performance	1.A lot of difficulty 2.Some difficulty 3.A little difficulty 4.No difficulty	Intensity	Preforming Vigorous activities	Running/ participating in sports	d Activities and participation (vigorous activities)	d4552 Running / d9201 Sports	Sports, vigorous
Item 2 of physical functioning	Walking as fast as others	Descriptive: capacity	1.A lot of difficulty 2.Some difficulty 3.A little difficulty 4.No difficulty	Intensity	Walking		d450 Walking		Fast as others
Item 3 of physical functioning	Carrying or lifting heavy things such as books, shopping, or school bags	Descriptive: Performance	1.A lot of difficulty 2.Some difficulty 3.A little difficulty 4.No difficulty	Intensity	lifting heavy objects		d4300 Lifting		
Item 4 of physical functioning	Climbing one flight of stairs	Descriptive: Performance	1.A lot of difficulty 2.Some difficulty 3.A little difficulty 4.No difficulty	Intensity	Climbing		d4551 Climbing		One flight of stairs

1. Name of instrument or other identifier	2. Verbatim health information (e.g., wording of item or instruction)	3. Perspective adopted in information	4. Response options	5. Classification of response options	6. Main concept: What is this information about?	7. Additional concepts contained in information	8. ICF category of main concept	9. ICF category of other concepts	10. Annotation
Item 5 of physical functioning	Climbing stairs as fast as others	Descriptive: capacity	1.A lot of difficulty 2.Some difficulty 3.A little difficulty 4.No difficulty	Intensity	Climbing		d4551 Climbing		Fast as others
Item 13 of physical functioning	To what extent do you have difficulty walking?	Appraisal	1.a long time without getting tired 2.a long time but get tired 4.cannot walk a long time because of getting tired quickly 5.avoid walking whenever possible because it's too tiring	Intensity	Walking		d450 Walking		
Item 19 of physical functioning	I have trouble recovering after physical effort	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Physical effort	Speed of recovery	nd-ph	nc	

1. Name of instrument or other identifier	2. Verbatim health information (e.g., wording of item or instruction)	3. Perspective adopted in information	4. Response options	5. Classification of response options	6. Main concept: What is this information about?	7. Additional concepts contained in information	8. ICF category of main concept	9. ICF category of other concepts	10. Annotation
Item 20 of physical functioning	I have to limit vigorous activities such as running or playing sports	Descriptive: Performance	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Limiting vigorous activities	Running/ participating in sports	d Activities and participation (vigorous activities)	d4552 Running / d9201 Sports	Sports, vigorous
Item 6 of vitality	You felt well	Appraisal	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel well		nd-gh		
Item 9 of vitality	You felt tired	Appraisal	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Tired		b1300 Energy level (tired)		
Item 10 of vitality	You felt full of energy	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel full energy		b1300 Energy level (full of energy)		
Item 11 of vitality	You felt exhausted	Appraisal	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel exhausted		b1300 Energy level (exhausted)		
Item 7 of emotional functioning	You felt worried	Appraisal	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel worried		b152 Emotional functions (worried)		

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Item 8 of emotional functioning	You felt useless	Appraisal	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel useless		b152 Emotional functions (useless)		
Item 12 of emotional functioning	You felt sad	Appraisal	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel sad		b152 Emotional functions (sad)		
Item 31 of emotional functioning	I often feel lonely	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	To feel lonely		b152 Emotional functions		
Item 33 of emotional functioning	It is difficult to make plans for the future (for example, going to college, getting married, getting promoted at work, etc.)	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Making plans for the future		b1641 Organization and planning		Going to college/getting married/getting promoted at work
Item 35 of role limitations	To what extent did you have trouble keeping up with your	Descriptive: Performance	1.You have had no trouble keeping up	Intensity	Work /school/ Regular daily activity		d850 Remunerative employmen		

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	schoolwork, professional work, or other daily activities during the past two weeks?		2.You have managed to keep up but it's been difficult 3.You have been behind 4.You have not been able to do these activities at all				t / d820 School education d230 Carrying out daily routine		
Item 36 of role limitations	How often were you absent from school, work, or unable to complete daily activities during the last two weeks because of your illness or treatments?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Frequency	Work /school/ Regular daily activity	Illness/treatment	d850 Remunerative employment / d820 School education d230 Carrying out daily routine	nd-hc	
Item 37 of role limitations	How often does CF get in the way of meeting your	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Frequency	School/work / personal goals	CF Illness	d850 Remunerative employmen	nd-hc	

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	school, work, or personal goals?						t / d820 School education		
Item 38 of role limitations	How often does CF interfere with getting out of the house to run errands such as shopping or going to the bank?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Frequency	Getting out of the house to run errands	CF Illness	d2 General tasks and demands	nd-hc	Shopping, going to the bank
Item 22 of social functioning	I have to stay at home more than I want to	Descriptive: Performance	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	To stay at home		nc		More than I want to
Item 23 of social functioning	I feel comfortable discussing my illness with others	Descriptive: Performance	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	To discuss illness with others		d355 Discussion		Feeling comfortable

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Item 27 of social functioning	People are afraid that I may be contagious	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	People are afraid of getting infected	Individuals' illness	e460 Societal attitudes	nd-hc	
Item 28 of social functioning	I get together with my friends a lot	Descriptive: Performance	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Getting together with friends		d7500 Informal relationships with friends		A lot
Item 29 of social functioning	I think my coughing bothers others	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Bothers others	Coughing	nc	nc	
Item 30 of social functioning	I feel comfortable going out at night	Descriptive: Performance	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Going out at night		nc		Feeling comfortable

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Item 40 of respiratory symptoms	Have you been congested?	Descriptive: Performance	1.A great deal 2.Somewhat 3.A little 4.Not at all	Intensity	Feeling congested		nc		
Item 41 of respiratory symptoms	Have you been coughing during the day?	Descriptive: Performance	1.A great deal 2.Somewhat 3.A little 4.Not at all	Intensity	Coughing		b4501 Functions of coughing		During the day
Item 42 of respiratory symptoms	Have you had to cough up mucus?	Descriptive: Performance	1.A great deal 2.Somewhat 3.A little 4.Not at all	Intensity	Coughing		b4501 Functions of coughing		Mucus
Item 43 of respiratory symptoms	Has your mucus been mostly:	Descriptive	1.clear 2.clear to yellow 3.Yellowish-green 4.green with traces of blood 5.don't know	Qualitative attributes	Mucus		nc		
Item 44 of respiratory symptoms	Have you been wheezing?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	To feel wheezing		b460 Sensations associated with cardiovascu		

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							lar and respiratory functions		
Item 45 of respiratory symptoms	Have you had trouble breathing?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Breathing		b440 Respiration functions		Trouble
Item 46 of respiratory symptoms	Have you woken up during the night because you were coughing?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Waking up at night	Coughing	b1342 Maintenance of sleep	b4501 Functions of coughing	
Item 14 of eating problems	How do you feel about eating?	Descriptive: Performance	1.Just thinking about food makes you feel sick 2.You never enjoy eating 3.You are sometimes able to enjoy eating 4.You are always able to enjoy eating	Qualitative attributes	To Feel about eating		b1302 Appetite		

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Item 21 of eating problems	I have to force myself to eat	Descriptive: Performance	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	To force oneself to eat		b1302 Appetite		
Item 50 of eating problems	Have you had eating problems?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Eating problems		b515 Digestive functions		
Item 24 of body image	I think I am too thin	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	To feel too thin		b1801 Body image		
Item 25 of body image	I think I look different from others my age	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	Look different from others same age		b1801 Body image		
Item 26 of body image	I feel bad about my physical appearance	Descriptive: Performance	1.Very true 2.Somewhat true	Intensity	Physical appearance		b1801 Body image		To feel bad

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			3.Somewhat false 4.Very false						
Item 47 of digestive symptoms	Have you had problems with wind?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Have problems with wind		b535 Sensations associated with the digestive system		
Item 48 of digestive symptoms	Have you had diarrhea?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Have diarrhea		b525 Defecation functions		
Item 49 of digestive symptoms	Have you had abdominal pain?	Descriptive: Performance	1.Always 2.Often 3.Sometimes 4.Never	Intensity	Have abdominal pain		b535 Sensations associated with the digestive system		
Item 39 of weight	Have you had trouble gaining weight?	Descriptive: Performance	1.A great deal 2.Somewhat 3.A little 4.Not at all	Intensity	Trouble gaining weight		b530 Weight maintenance functions		
Item 15 of treatment burden	To what extent do your treatments make your daily life more difficult?	Descriptive: Performance	1.Not at all 2.A little 3.Moderately 4.A lot	Intensity	Treatments	Daily life	d570 Looking after one's health	d230 Carrying out daily routine	

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Item 16 of treatment burden	How much time do you currently spend each day on your treatments?	Descriptive: Performance	1.A lot 2.Some 3.A little 4.Not very much	Duration	Time spent daily on treatment		d570 Looking after one's health		
Item 17 of treatment burden	How difficult is it for you to do your treatments (including medications) each day?	Descriptive: Performance	1.Not at all 2.A little 3.Moderately 4.A lot	Intensity	Difficulty of doing daily treatments		d570 Looking after one's health		
Item 18 of general health perception	How do you think your health is now?	Appraisal	1. Excellent 2. Good 3. Fair 4. Poor	Intensity	Health		nd-gh		
Item 32 of general health perception	I feel healthy	Appraisal	1.Very true 2.Somewhat true 3.Somewhat false 4.Very false	Intensity	To feel healthy		nd-gh		
Item 34 of general health perception	I lead a normal life	Appraisal	1.Very true 2.Somewhat true	Intensity	Normal life		nc		

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			3.Somewhat false 4.Very false						
Borg scale (rate of perceived exertion)[129,130]									
	Where would you rate the difficulty of your breathing on this scale	Appraisal	0 Nothing at all 0.5 Very, very slight 1 Very slight 2 Slight 3 Moderate 4 Somewhat severe 5 Severe 6 7 Very severe 8 9 Very, very severe 10 Maximal shortness of breath	Intensity	difficulty of breathing		b460 Sensations associated with cardiovascular and respiratory functions		
	Where would you rate the fatigue of your lower extremity on this scale	Appraisal	0 Nothing at all 0.5 Very, very slight 1 Very slight 2 Slight 3 Moderate 4 Somewhat severe 5 Severe 6 7 Very severe 8 9 Very, very	Intensity	Fatigue of lower extremity		b740 muscle endurance functions		

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			severe 10 Maximal fatigue						
Visual analogue scale (VAS)[131]									
	Where would you put your pain on this scale	Appraisal	From 0 to 10 0 being the least possible pain and 10 the worst possible pain	Intensity	Pain		b280 Pain		
Hospital Anxiety and Depression Scale (HADS)[132]									
Item 1	I feel tense or 'wound up'	Appraisal	1.Most of the time 2.A lot of the time 3.From time to time, occasionally 4.Not at all	Intensity	Feel tense or 'wound up'		b1521 Regulation of emotion		
Item 2	I still enjoy the things I used to enjoy	Appraisal	1.Definitely as much 2.Not quite so much 3.Only a little 4.Hardly at all	Intensity	enjoy the things as usual		b1522 Range of emotion		

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Item 3	I get a sort of frightened feeling as if something awful is about to happen	Appraisal	1.Very definitely and quite badly 2.Yes, but not too badly 3.A little, but it doesn't worry me 4.Not at all	Intensity	To get frightened feeling		b1522 Range of emotion		As if something awful is about to happen
Item 4	I can laugh and see the funny side of things	Appraisal	1.As much as I always could 2.Not quite so much now 3.Definitely not so much now 4.Not at all	Intensity	Laughing		b1520 Appropriateness of emotion		See the funny side of things
Item 5	Worrying thoughts go through my mind	Appraisal	1.A great deal of the time 2.A lot of the time 3.From time to time but not too often 4.Only occasionally	Intensity	Worrying thoughts		b1263 Psychic stability		

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Item 6	I feel cheerful	Appraisal	1.Not at all 2. Not often 3.Sometimes 4.Most of the time	Intensity	To feel cheerful		b1265 Optimism		
Item 7	I can sit at ease and feel relaxed	Appraisal	1.Definitely 2.Usually 3.Not often 4.Not at all	Intensity	To feel relaxed		b1263 Psychic stability		
Item 8	I feel as if I am slowed down	Appraisal	1.Nearly all the time 2.Very often 3.Sometimes 4.Not at all	Intensity	To feel slowed down		b1300 Energy level		
Item 9	I get a sort of frightened feeling like 'butterflies' in the stomach	Appraisal	1.Not at all 2.Occasionally 3.Quite often 4.Very often	Intensity	To get frightened feeling		b1522 Range of emotion		Like 'butterflies' in the stomach
Item 10	I have lost interest in my appearance	Appraisal	1.Definitely 2.I don't take so much care as I should	Intensity	To lose interest in appearance		b180 Experience of self and time functions		

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			3.I may not take quite as much care 4.I take just as much care as ever						
Item 11	I feel restless as if I have to be on the move	Descriptive: Performance	1.Very much indeed 2.Quite a lot 3.Not very much 4.Not at all	Intensity	To feel restless		b1263 Psychic stability		As if I have to be on the move
Item 12	I look forward with enjoyment to things	Appraisal	1.As much as I ever did 2.Rather less than I used to 3.Definitely less than I used to 4.Hardly at all	Intensity	Looking forward with enjoyment to things		b1265 Optimism		
Item 13	I get sudden feelings of panic	Appraisal	1.Very often indeed 2.Quite often 3.Not very often 4.Not at all	Intensity	To get sudden feelings of panic		b152 Emotional functions		
Item 14	I can enjoy a good book or	Appraisal	1.Often 2.Sometimes 3.Not often	Intensity	To enjoy things		b1522 Range of emotion		A good book or

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	radio or TV program		4.Very seldom						radio or TV program
Fatigue Inventory[133]									
	Fatigue severity items	Appraisal	I feel tired Physically, I feel exhausted I feel fit I feel weak I feel rested Physically I am in bad shape I tire easily Physically I feel I am in good shape	Qualitative attributes	General fatigue		b4552 Fatiguability		
	Activity items	Descriptive: Performance	I think I do a lot in a day I think I do very little in a day I get little done	Qualitative attributes	Activity of daily living		d230 Carrying out daily routine	b1300 Energy level	
	Concentration items	Descriptive: Performance	Thinking requires effort When I am doing something, I can keep my thoughts on it	Qualitative attributes	Concentration		b140 Attention functions		

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			I find it easy to concentrate It takes a lot of effort to concentrate on things My thoughts easily wander						
	Motivation items	Descriptive: Performance	I feel very active I feel like doing all kinds of nice things I have a lot of plans	Qualitative attributes	Motivation		b1301 Motivation		
Physical activity recall questionnaires[134]									
Item 1	On the average, how many hours did you sleep each night during the last five weekday nights (Sunday-Thursday)?	Appraisal	____hours	Duration	Hours of sleep each night		b1340 Amount of sleep		During the last five weekday nights (Sunday-Thursday)
Item 2	On the average, how many hours did you sleep each night last	Appraisal	____hours	Intensity	Hours of sleep each night		b1340 Amount of sleep		Last Friday and Saturday nights

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	Friday and Saturday nights?								
Item 3	Consider moderate activities. What activities did you do and how many total hours did you spend during the last 5 weekdays doing these moderate activities or others like them?	Appraisal	Please tell me to the nearest half hour. — hours	Duration	Hours spent doing moderate activities		d5701 Managing diet and fitness		During the last 5 weekdays
Item 4	Last Saturday and Sunday, how many hours did you spend on moderate activities and what did you do? (Probe: Can you think of any other sports, job, or household activities that	Appraisal	____hours	Duration	Hours spent doing moderate activities		d5701 Managing diet and fitness		Can you think of any other sports, job, or household activities that would fit into this category?

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	would fit into this category?)								
Item 5	hard activities. What activities did you do and how many total hours did you spend during the last 5 weekdays doing these hard activities or others like them?	Appraisal	Please tell me to the nearest half hour — hours	Duration	Hours spent doing hard activities		d5701 Managing diet and fitness		During the last 5 weekdays
Item 6	Last Saturday and Sunday, how many hours did you spend on hard activities and what did you do? (Probe: Can you think of any other sports, job, or household activities that would fit into this category?)	Appraisal	____hours	Duration	Hours spent doing hard activities		d5701 Managing diet and fitness		Can you think of any other sports, job, or household activities that would fit into this category?

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Item 7	very hard activities. What activities did you do and how many total hours did you spend during the last 5 weekdays doing these very hard activities or total hours did you spend during the last 5 weekdays doing these very hard activities or others like them?	Appraisal	Please tell me to the nearest half hour. - hours	Duration	Hours spent doing very hard activities		d5701 Managing diet and fitness		During the last 5 weekdays
Item 8	Last Saturday and Sunday, how many hours did you spend on very hard activities and what did you do? (Probe: Can you think of any other sports, job, or household	Appraisal	____hours	Duration	Hours spent doing very hard activities		d5701 Managing diet and fitness		Can you think of any other sports, job, or household activities that would fit into this category?

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	activities that would fit into this category?)								
Item 9	Compared with your physical activity over the past 3 months, was last week's physical activity more, less, or about the same?	Appraisal	1. More 2. Less 3. About the same	Intensity	Physical activity		d5701 Managing diet and fitness		
The Quality of Well-Being Scale[135,136]									
	Mobility scale	Descriptive: Performance	5. No limitations for health reasons 4. Did not drive a car, health related: did not ride in a car as usual for age (15 yr) (health related), and /or did not use public transportation (health related), or had or would	Intensity	Mobility		d4 Mobility d47-d489 Moving around using transportation		Health related

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			have used more help than usual for age to use public transportation (health related) 2. In hospital, health related						
	Physical activity scale	Descriptive: Performance	4. No limitations for health reasons 3. In wheelchair, moved of controlled movement of wheelchair without help from someone else, or had trouble or did not try to lift, stoop, bend over, or use stairs or inclines (health related) and /or limped, used a cane,	Intensity	Physical activity		d4 Mobility d5701/Managing diet and fitness		Health related

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			crutches, or walker (health related), and /or had any other physical limitation in walking, or did not try to walk as far or as fast as others the same age are able (health related) 1. In wheelchair, did not move or control the movement of wheelchair without help from someone else, or in bed, chair, or couch for most or all of the day (health related)						
	Social activity scale	Descriptive: Performance	5. No limitations for health reasons	Intensity	Social activity		d920 Recreation and		health related

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			4. Limited in other (e.g., recreational) role activity (health related) 3. Limited in major (primary) role activity (health related) 2. Performed no major role activity (health related) but did Preform self-care 1. Performed no major role (health related) and did not perform or had more trouble than usual in performance of one or more self-care activities (health related)				leisure/d230 Carrying out daily routine activities/d570 Looking after one's health		