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ROLE OF PHYSICAL ACTIVITY IN THE MULTIDISCIPLINARY MANAGEMENT OF CYSTIC FIBROSIS: A LITERATURE REVIEW

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ABSTRACT

Introduction: Cystic fibrosis (CF) is a multisystem genetic disorder in which progressive pulmonary disease remains the leading cause of morbidity and mortality. Advances in pharmacological therapy, particularly CFTR modulators, have transformed the clinical landscape; however, non-pharmacological strategies—especially structured physical activity (PA)—remain essential for optimising long-term outcomes. This review synthesises current evidence on the physiological basis, clinical benefits, safety considerations, and practical implementation of PA as part of multidisciplinary CF management.
Methods: A comprehensive literature search was conducted in PubMed, Scopus, and Web of Science for peer-reviewed articles published between 2020 and 2025. Search terms included cystic fibrosis; physical activity; exercise training; pulmonary rehabilitation; multidisciplinary care. Randomised controlled trials, systematic reviews, meta-analyses, and relevant clinical guidelines were included.
Results: Evidence consistently shows that regular physical activity improves exercise capacity, enhances mucus clearance, and contributes to stabilizing or improving selected parameters of lung function in people with cystic fibrosis. Aerobic, endurance, combined, and inspiratory muscle training provide complementary benefits for cardiovascular fitness, muscle strength, bone health, and mental well-being. Home-based programs, supported by telemedicine, offer comparable results to in-person training while reducing the risk of infection. Regular physical activity is often hindered by disease symptoms, limited access to appropriate sports facilities, and psychosocial limitations. However, there are facilitating factors, such as personalized exercise recommendations, support from an interdisciplinary team, and digital monitoring technologies, which enable ongoing patient engagement.
Conclusions: Physical activity is a safe, effective, and versatile component of multidisciplinary CF care, offering physiological, functional, and psychosocial benefits that complement pharmacological and airway clearance therapies. Integrating tailored exercise interventions into routine management should be prioritised to maximise health outcomes and quality of life in this population.

KEYWORDS
Cystic Fibrosis, Physical Activity, Exercise Training, Pulmonary Rehabilitation, Multidisciplinary Care

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Pathophysiology, clinical manifestations and treatment of cystic fibrosis

Pathophysiology

Cystic fibrosis (CF) is an autosomal recessive disease caused by mutations in the *CFTR* gene, resulting in defective chloride and bicarbonate ion transport across epithelial surfaces. This dysfunction leads to the production of abnormally thick, viscous mucus, predominantly affecting the lungs, pancreas, hepatobiliary tract, and intestines. In the respiratory system, this mucus impairs mucociliary clearance, fostering persistent bacterial infections and neutrophilic inflammation. Over time, this results in bronchiectasis and irreversible decline in pulmonary function [1, 2].

Clinical Features

CF typically presents in early childhood, yet progression varies based on genotype. Respiratory symptoms include chronic productive cough, frequent exacerbations, wheezing, and progressive respiratory failure. Exocrine pancreatic insufficiency contributes to malabsorption, fat-soluble vitamin deficiencies, and poor growth. Elevated sweat chlorine levels remain diagnostic benchmarks. Other signs may include chronic sinusitis, nasal polyps, digital clubbing, and male infertility, often due to congenital bilateral absence of the vas deferens (CBAVD) [3, 4].

Treatment of Cystic Fibrosis

1. Pharmacological Management

CFTR modulator therapies, particularly the triple combination of elexacaftor/tezacaftor/ivacaftor, have revolutionised CF treatment, demonstrating marked improvements in lung function, nutritional parameters, and quality of life in both clinical trials and real-world settings [5, 6]. The emergence of next-generation CFTR modulators—such as vanzacaftor/tezacaftor/deutivacaftor and other novel triple combinations currently in late-phase clinical trials—has broadened therapeutic options for patients with both common and rare CFTR mutations [7, 8]. These advancements enable precision medicine approaches, where treatments are tailored to an individual's mutation profile and clinical phenotype, as recommended by the 2023 European Cystic Fibrosis Society Standards [9] and NIH guidance [10].

2. Non-pharmacological Management

Non-pharmacological interventions are an important complement to pharmacotherapy in the treatment of cystic fibrosis (CF), helping to preserve lung function and improve the patient's well-being and nutritional status.

- Airway Clearance Techniques (ACTs).

Methods such as active cycle of breathing, positive expiratory pressure, chest wall oscillation, and autogenic drainage facilitate mucus clearance and mitigate infection risk. Individualisation based on age, disease severity, and patient preference is recommended to support long-term adherence [11, 12].

- Nutritional Management and Pancreatic Enzyme Replacement Therapy (PERT).

Maintaining nutritional adequacy is essential—for respiratory muscle function and quality of life. Guideline-based PERT doses, high-calorie diet plans, and fat-soluble vitamin supplementation are crucial for individuals with exocrine pancreatic insufficiency. Recent studies emphasise regular monitoring and dose adjustments to ensure effective nutrient absorption [13].

- Psychosocial Support.

Psychological interventions—such as counselling, cognitive-behavioural strategies, and family-centred education—have been shown to improve adherence to inhaled therapies in individuals with CF, without contributing to increased anxiety or depression [14]. Additionally, evolving psychosocial stressors necessitate responsive care models that are flexible and patient-centred [15].

- Telehealth and Remote Monitoring.

Telemedicine platforms now enable remote supervision of ACTs, multidisciplinary consultations, and monitoring of treatment adherence. Hybrid models that combine in-person evaluations with virtual follow-ups are considered practical and well accepted while reducing cross-infection risk [16].

Regular physical activity and structured exercise remain key components of CF treatment as they improve functional capacity and well-being – an aspect that will be discussed in the next section.

Role of Physical Activity in Cystic Fibrosis Management

Physiological Basis

In cystic fibrosis (CF), the accumulation of dehydrated, viscous mucus within the airways results from impaired CFTR-mediated ion transport, creating a cycle of mucus stasis, bacterial colonisation, and chronic neutrophilic inflammation [1, 2]. Physical activity (PA) exerts multiple physiological effects that directly address these pathophysiological processes. Increased tidal volume and minute ventilation during exercise enhance shear forces along the bronchial walls, facilitating mucus mobilisation and clearance [12]. Repeated bouts of activity also stimulate surfactant production, improving airway patency. Beyond pulmonary effects, PA induces cardiovascular adaptations—including increased stroke volume, improved peripheral oxygen extraction, and capillary density—that reduce ventilatory demand for a given workload [17]. Skeletal muscle adaptations, particularly increased oxidative enzyme activity and mitochondrial density, enhance endurance and reduce peripheral fatigue [18]. Collectively, these effects help interrupt the cycle of infection, inflammation, and declining lung function in CF.

Impact on Exercise Capacity

Exercise capacity in CF is typically reduced due to ventilatory limitation, muscle deconditioning, and systemic inflammation [19]. Peak oxygen uptake (VO_2peak), an established prognostic marker, is often 20–40% lower in CF patients compared to healthy peers [20]. Longitudinal studies demonstrate that structured aerobic training can increase VO_2peak by 10–20% over 8–12 weeks [21]. Other key performance indicators—such as ventilatory threshold (VT), minute ventilation (VE), oxygen pulse (O_2 pulse), and six-minute walk distance (6MWD)—also improve with sustained training [22]. Interval-based and combined aerobic–resistance protocols appear particularly effective in enhancing both central (cardiovascular) and peripheral (muscular) components of exercise capacity [23]. Notably, improvements in exercise tolerance are strongly correlated with better health-related quality of life (HRQoL) scores and reduced hospitalisation rates [24].

Impact on Pulmonary Function

While the impact of PA on forced expiratory volume in one second (FEV_1) is variable—likely due to differences in disease stage, intervention intensity, and adherence—emerging evidence supports beneficial effects on other pulmonary parameters [25]. Aerobic and combined exercise programmes have been associated with stabilisation or modest improvements in forced vital capacity (FVC), forced expiratory flow at 25–75% of FVC (FEF25–75), and mid-expiratory flow rates (MEF50) [26]. Reductions in residual volume to total lung capacity ratio (RV/TLC) have been reported, suggesting alleviation of air trapping [27]. Additionally, exercise may improve lung clearance index (LCI), derived from multiple-breath washout testing, which is more sensitive than spirometry for detecting early small airway disease [28]. These physiological benefits likely result from enhanced mucus clearance, reduced airway inflammation, and improved respiratory muscle performance.

Adjunctive Benefits and Clinical Implications

Regular PA confers systemic benefits beyond the respiratory system. Exercise promotes bone mineral density—particularly relevant in CF, where osteopenia is prevalent—via mechanical loading and endocrine stimulation [29]. It also improves insulin sensitivity, potentially reducing the risk or severity of cystic fibrosis-related diabetes (CFRD) [30]. Importantly, psychological benefits such as reduced anxiety and improved mood contribute to better treatment adherence and overall wellbeing [14]. These multidimensional effects support the integration of structured PA into standard CF care, alongside pharmacological therapy, nutritional management, and airway clearance techniques.

Types of Exercise Interventions in Cystic Fibrosis

Aerobic Training

Aerobic training remains a cornerstone of physical activity interventions in cystic fibrosis (CF), aiming to enhance cardiovascular efficiency, improve oxygen transport, and facilitate mucus clearance. Typical modalities include cycling, treadmill walking or running, and swimming. Training intensities are often prescribed using a percentage of peak oxygen uptake (VO_2peak) or heart rate reserve, with evidence suggesting that moderate-to-vigorous intensity sustained for 20–40 minutes, three to five times per week, yields significant improvements in VO_2peak, ventilatory threshold, and six-minute walk distance (6MWD) [31, 32]. Beyond cardiorespiratory benefits, aerobic exercise is associated with improved lung clearance index (LCI) and reduced daily symptom burden [33].

Resistance Training

Resistance training targets muscle strength and endurance, addressing the muscle wasting and weakness frequently observed in CF due to chronic inflammation, malnutrition, and corticosteroid exposure. Programmes typically incorporate multi-joint exercises (e.g., squats, lunges, push-ups) or resistance machines, with intensity guided by repetition maximum (RM) principles. Studies indicate that two to three weekly sessions over 8–12 weeks can improve isometric and dynamic muscle strength, enhance functional performance, and support bone mineral density maintenance [34, 35]. Gains in muscular strength may indirectly improve ventilatory efficiency by reducing the relative load on respiratory muscles during physical activity [36].

Combined Training

Combined aerobic–resistance protocols appear to offer synergistic benefits, simultaneously enhancing cardiovascular fitness and muscular performance. Randomised trials have reported that mixed training regimens can increase VO_2peak by 10–15% while also improving peripheral muscle strength and endurance [37]. These programmes may also promote greater adherence, as variety in exercise modalities reduces monotony and allows for adaptation to fluctuating health status [38].

Inspiratory Muscle Training (IMT)

IMT is a method for improving the strength and endurance of the respiratory muscles, particularly the diaphragm and intercostal muscles. Using devices measuring threshold load, patients perform repetitive inspiratory efforts against resistance, typically 5 to 7 days per week for 15–30 minutes per day. Meta-analyses suggest that IMT can improve maximum inspiratory pressure (MIP), inspiratory muscle endurance, and perceived dyspnea, although its effect on FEV_1 remains inconclusive [39, 40].

Home-based and Telehealth-Supported Exercise

Advances in digital health have facilitated the delivery of supervised exercise interventions beyond the clinic setting. Home-based programmes, supported by telehealth consultations, video demonstrations, and wearable activity trackers, have demonstrated comparable improvements in exercise capacity and quality of life to centre-based training [42]. Remote supervision reduces cross-infection risks, enhances accessibility for patients in geographically remote areas, and supports continuity during acute illness or hospitalisation [43]. Hybrid models—blending in-person baseline assessments with ongoing virtual follow-up—are increasingly recognised as a practical and effective approach for long-term exercise adherence [44].

Barriers and Facilitators to Physical Activity in Cystic Fibrosis

Barriers

Engagement in regular physical activity (PA) among individuals with cystic fibrosis (CF) is influenced by a combination of physiological, psychological, and environmental factors.

Physiologically, disease-related symptoms such as chronic cough, dyspnoea, fatigue, and musculoskeletal pain can limit exercise tolerance and discourage participation [45, 46]. Pulmonary exacerbations, often requiring prolonged rest or hospitalisation, may disrupt established exercise routines and lead to detraining [47].

Psychological barriers include low self-efficacy, anxiety related to symptom exacerbation during exercise, and reduced motivation—particularly in adolescents and adults balancing high treatment burdens with educational or occupational demands [48]. Social and environmental constraints, such as limited access to suitable exercise facilities, infection control precautions, and lack of specialised supervision, further restrict opportunities for safe participation [49].

Facilitators

Conversely, several factors can enhance PA engagement in CF. Individualised exercise prescriptions that align with personal preferences and symptom patterns have been shown to improve adherence [50]. Multidisciplinary team involvement—including physiotherapists, exercise physiologists, and psychologists—provides the necessary clinical oversight, education, and motivational support [51]. Technological solutions, such as wearable activity trackers and telehealth-based coaching, facilitate remote monitoring and feedback, enabling continuous engagement even during illness or travel restrictions [52]. Social support from family members and peers, alongside integration of PA into routine daily activities, reinforces long-term behaviour change [53]. Emerging evidence also highlights the role of positive exercise experiences in enhancing quality of life and fostering intrinsic motivation [54].

Clinical Safety Considerations for Exercise in Cystic Fibrosis

Exercise is generally safe and beneficial for individuals with cystic fibrosis (CF), but it requires careful planning and monitoring due to disease-specific risks. Awareness of potential complications and appropriate safety measures ensures that physical activity remains a supportive, rather than detrimental, component of multidisciplinary care.

Airway Obstruction and Secretion Mobilisation

The increased ventilation during exercise can mobilise airway secretions, improving mucus clearance but occasionally precipitating transient airway obstruction or bronchospasm, particularly in those with advanced lung disease [55, 56]. Pre-exercise use of bronchodilators and incorporating airway clearance techniques before and after sessions can mitigate these risks.

Hypoxemia

Patients with moderate to severe pulmonary impairment are at risk of exercise-induced desaturation (SpO_2 <90%), particularly during high-intensity or prolonged activity. Pulse oximetry monitoring during initial sessions helps identify those requiring supplemental oxygen or intensity adjustments. Targeting submaximal workloads that maintain adequate oxygenation is recommended for high-risk individuals [57].

Dehydration and Electrolyte Imbalance

Due to elevated sweat sodium and chloride losses, CF patients are prone to dehydration and electrolyte disturbances, especially in hot or humid environments. Adequate fluid and salt replacement—tailored to exercise duration, environmental conditions, and individual sweat rates—should be an integral part of exercise planning [58].

Hemoptysis

Mild hemoptysis is not uncommon in advanced CF and may be exacerbated by strenuous exercise through increased intrathoracic pressure and vascular fragility. Significant or recurrent bleeding warrants temporary cessation of high-intensity activity and prompt medical evaluation. Low-intensity training may be resumed once the episode resolves and underlying causes are addressed [59].

Monitoring and Programme Modification

Exercise prescription in CF should follow a personalised, stepwise approach based on baseline lung function, comorbidities, and prior activity history [60]. Initial sessions should be supervised—preferably by physiotherapists experienced in CF—allowing for adjustments in intensity, duration, and modality according to symptom response, oxygenation, and fatigue levels. Periodic reassessment of pulmonary function, exercise tolerance, and patient-reported symptoms ensures ongoing safety and efficacy [61]. Telehealth-enabled monitoring can extend this oversight to home-based training programmes, enhancing adherence while maintaining safety [16].

By integrating safety protocols into exercise prescription, clinicians can maximise benefits while minimising risks, ensuring that physical activity remains a sustainable and effective component of CF care.

Practical Recommendations for Physical Activity in Cystic Fibrosis – Summary

	Recommendation	Practical Notes	References
Frequency	3–5 days/week of structured physical activity	Encourage daily incidental activity to reduce sedentary time	[63]
Intensity	Moderate-to-vigorous (e.g., interval training)	High-intensity sessions can improve exercise capacity and muscle strength in CF patients	[64]
Type	Combination of aerobic, resistance, and inspiratory muscle training	Aerobic: improves cardiovascular fitness and mucus clearance; Resistance: supports muscle and bone health	[64, 65]
Time / Duration	20–60 min per session	Can be split into shorter bouts; adjust during exacerbations	[64]
Integration with ACTs	Exercise before or after airway clearance techniques	Pre-exercise bronchodilator may reduce bronchospasm; exercise supports mucus clearance	[66]
Monitoring / Safety	Monitor SpO_2, heart rate, RPE, hydration, and signs of overexertion	Keep $SpO_2 > 90\%$; watch for dyspnea, fatigue, hemoptysis	[63, 64]
Individualization & Progression	Adapt to age, lung function, comorbidities, nutritional status; gradual progression	Combine home-based and supervised training for optimal adherence	[62]
Behavioral Strategies	Goal-setting, self-monitoring, family support, telehealth interventions	Personalized approach and remote support improve engagement	[65, 66]
Special Considerations	Modify training during exacerbations, acute illness, or treatment changes	Promote lifelong engagement; adapt to evolving therapies (e.g., CFTR modulators)	[63, 66]

Conclusions

Cystic fibrosis (CF) remains a progressive, life-limiting disease in which pharmacological advances, particularly CFTR modulators, have dramatically improved survival and quality of life. Nevertheless, non-pharmacological interventions—most notably structured physical activity (PA)—are indispensable components of comprehensive care. Current evidence demonstrates that regular PA enhances exercise capacity, contributes to airway clearance, supports pulmonary function, and provides systemic benefits for musculoskeletal health, metabolic regulation, and psychological well-being.

Importantly, PA is safe across disease stages when delivered with appropriate monitoring and individualisation. Exercise interventions encompassing aerobic, resistance, and inspiratory muscle training should be integrated into routine management, with flexibility to accommodate disease fluctuations and patient preferences. The expansion of telehealth and home-based programmes has created new opportunities for sustained engagement, while multidisciplinary support and personalised exercise prescriptions remain key to overcoming adherence barriers.

Future research should focus on optimising exercise protocols, identifying long-term adherence strategies, and exploring the interaction between PA and novel pharmacotherapies. From a clinical perspective, structured PA should be promoted not as an optional adjunct, but as a fundamental pillar of multidisciplinary CF care—essential for maximising health outcomes, functional independence, and overall quality of life.

Disclosure
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Methodology: Diana Wisz, Filip Maciej Huzarski, Katarzyna Maria Turek,
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