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A Breath Full of Life – The Importance of Movement and Physiotherapy in the Treatment of Children with Cystic Fibrosis

Katarzyna Oświeczyńska

Silesian Academy, Zbigniew Religa Faculty of Medical Sciences

E-mail: kasia.igonek@gmail.com

https://orcid.org/0009-0000-8383-9175

Aleksandra Sowa

Medical University of Wroclaw

E mail: sowaolaa@gmail.com

https://orcid.org/0009-0009-3606-8756

Kacper Trząsalski

Medical University of Wroclaw

E-mail: kacper.trzasalski1@gmail.com

https://orcid.org/0009-0002-8425-6865

Patrycja Jędrzejewska-Rzezak. M.D.
The John Paul II Catholic University of Lublin
E-mail: patrycjaj@kul.lublin.pl
https://orcid.org/0000-0003-2144-5810

Abstract

Cystic fibrosis is a congenital genetic disease that cannot be acquired or passed on through contact with another person. It occurs in both boys and girls because its cause is a genetic mutation present from birth. The disease develops when a child inherits two defective genes - one from the mother and the other from the father. Parents, although not suffering from the disease themselves, are carriers of the mutation and can pass it on to their offspring. Chronic inflammation gradually destroys the lung parenchyma, causing changes such as bronchodilation, fibrosis and sometimes emphysema or atelectasis. As the disease progresses, respiratory failure occurs, leading to hypoxia and a significant reduction in physical endurance in patients with cystic fibrosis. Regular physical activity and properly selected physiotherapy are a key component of cystic fibrosis therapy. They support the functioning of the respiratory system, improving patients' quality of life. Systematic exercise and specialized drainage techniques facilitate the removal of secretions from the lungs, strengthen respiratory muscles and increase overall fitness.

Aim of the study: The purpose of this paper is to provide a detailed insight into cystic fibrosis as a disease entity and to analyze the use of various forms of exercise, physiotherapy and adaptation of breathing exercises in children suffering from this disease, based on a review of available studies and literature.

Materials and methods: A review of the literature available in the PubMed and Google Scholar database was performed, using the key words: “cystic fibrosis”, „genetic disease”, „CFTR gene”, „respiratory physiotherapy”, “physical activity”, “respiratory system”, “breathing exercises”.

Keywords: cystic fibrosis, genetic disease, CFTR gene, respiratory physiotherapy, physical activity, respiratory system, breathing exercises

1. Introduction

Cystic fibrosis (CF) is a congenital genetic disease caused by a mutation of the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene located on chromosome 7. It is inherited in an autosomal recessive manner according to Mendel's laws, meaning that symptoms appear only in people who have inherited a mutated version of the gene from both parents [1].

The CFTR gene is responsible for producing a protein that regulates the transport of chloride ions across cell membranes. Defects in its structure or function lead to dysfunction of chloride channels, resulting in excessive mucus thickening in the body. Thick and sticky mucus accumulates mainly in the respiratory tract and exocrine glands, causing chronic lung infections, pancreatic insufficiency, liver disease and reduced fertility.

Cystic fibrosis is most common in people of European descent [2] .

To date, more than 1,500 mutations in the CFTR gene have been identified, the most common of which - ΔF508 - affects about 70% of patients [3] . The disease affects many systems, particularly the respiratory and digestive systems, leading to metabolic problems and difficulties with nutrient absorption.

Regular exercise plays a serious role in the treatment of cystic fibrosis, facilitating the elimination of accumulated mucus from the airways, strengthening respiratory muscles and increasing overall physical fitness, improving the patient's condition. This article focuses on the impact of exercises to support lung function in children with cystic fibrosis and presents the most effective examples of exercises to support the breathing process.

2. Cystic fibrosis respiratory symptoms

Cystic fibrosis is a condition that impacts many systems; nonetheless, the primary cause of morbidity and mortality in affected individuals is worsening lung illness. Stagnation of mucous discharges is characteristic of CF. Effective mucociliary clearance is impeded by the limited volume of periciliary fluid in the respiratory tract epithelium. The alteration in pH of the surface fluid resulting from elevated NaCl concentration leads to a reduction in the efficacy of antibacterial agents, hence facilitating various infections. *Staphylococcus aureus* and *Haemophilus influenzae* are prevalent bacteria responsible for respiratory tract infections in early childhood; however, *Pseudomonas aeruginosa* is predominantly isolated from

respiratory secretions in the majority of patients [4][5]. Bacteria typically establish a biofilm, which serves as a formidable barrier against phagocytic cells and pharmaceuticals. Persistent active infections, characterized by the accumulation of inflammatory cells and the discharge of their constituents, result in damage to the bronchial walls, leading to a loss of cartilage support and muscle tone, ultimately causing bronchiectasis [6][7].

Respiratory clinical manifestations of CF typically commence in early childhood with a recurring cough that evolves into a chronic condition as the disease advances. A productive cough ultimately affects patients on a daily basis. Disease progression is marked by abrupt exacerbations featuring heightened cough, accelerated respiration, dyspnea, and augmented sputum output. These episodes result in acute, transient impairment of lung function, which improves with suitable therapy; nonetheless, the progression of the disease leads to irreversible decline in lung function and progressive respiratory failure. In patients with CF, an increase in the anteroposterior width of the chest is noted due to air trapping resulting from prolonged airway blockage caused by thick mucus accumulation. Hyperinflation frequently arises from airway obstruction. Airway hyperresponsiveness, clinically evident as wheezing, is another symptom observed in patients with CF. Individuals with severe pulmonary illness may experience spontaneous pneumothorax and hemoptysis. A serious consequence of chronic lung disease CF is pulmonary hypertension, which correlates with markedly reduced survival rates.

The majority of patients with CF develop chronic sinusitis as a result of poor clearance of viscous secretions from the sinuses. Subsequent structural damage may ensue. Symptomatic polyps may develop in the sinuses over time. Children have persistent nasal obstruction and debilitating headaches, which impair their overall functioning [8].

3. Fundamental management for individuals with cystic fibrosis

Cystic fibrosis is a chronic condition necessitating interdisciplinary management, including a pulmonologist, physiotherapist, dietitian, psychologist, and other specialists. The paramount aspect of chronic treatment is the systematic facilitation of clearing retained secretions from the respiratory tract, ensuring adequate nutrition for the patient, preventing and effectively managing bacterial infections, mitigating inflammation, and preventing complications while administering appropriate treatment if they arise. In the majority of cystic fibrosis patients, there exists the potential for causative treatment utilizing medications that precisely alter the

CFTR protein. Lung transplantation serves as a treatment option for severely advanced lung disease [9][10].

It is crucial to eliminate accumulated thick secretions from the airways. It should be executed promptly following the diagnosis of CF. Respiratory physiotherapy must be conducted daily for the entirety of the patient's life, comprising two or more sessions each day. A variety of airway clearance strategies are presently accessible, and the selection of methods ought to be customized to the specific requirements of the patient. Conventional drainage techniques encompass postural drainage, percussion, and chest shaking [11] . Efficient coughing and forced exhalation techniques are beneficial. Patients have access to basic equipment that facilitates bronchial drainage, including Flutter, Acapella, and a PEP mask. Effectively administered rehabilitation enhances the volume of expectorated secretions, diminishes symptom severity, decreases the frequency of exacerbations, and so improves pulmonary function while impeding disease progression [12][13] [14].

In individuals with CF, an appropriate diet is of paramount significance. A high-fat diet supplemented with fat-soluble vitamins is advised due to the concurrent exocrine pancreatic insufficiency. Furthermore, individuals with CF ought to ingest high-calorie and high-protein meals to sustain an appropriate body weight. It is frequently essential to augment the diet with enzyme supplements. The Cystic Fibrosis Foundation currently recommends that women with cystic fibrosis maintain a daily caloric intake of 2,500 to 3,000 calories, and men with the condition should consume between 3,000 and 3,700 calories daily. Oral alimentation is favored. Enteral nutrition should be considered solely when intake fails to satisfy metabolic demands, resulting in a progressive decline in body mass index. Parenteral feeding should be considered solely when oral or enteral nutrition fails to fulfill metabolic requirements [15].

Chronic mucolytic therapy, administered via inhalation, is utilized with respiratory physiotherapy and nutritional management. To expedite and enhance the clearance of secretions from the respiratory tract, inhalation of dornase alfa (at least 30 minutes prior to drainage) or inhalation of hypertonic saline solution (7%) is employed. Dornase alfa, when administered continuously, decreases the incidence of exacerbations and enhances the overall quality of life for those with CF [16][17]. It is important to note that bronchodilators must be taken prior to the inhalation of concentrated salt. In individuals who are intolerant to or inadequately respond to inhalation of dornase or concentrated NaCl, mannitol inhalation may be employed as a second-line therapy.

It is crucial to note the latest class of pharmaceuticals accessible for CF patients, specifically CFTR modulators. The advent of these medications provides patients with

optimism for an extended life in robust health. CFTR modulators are oral pharmaceuticals that enhance the synthesis, intracellular processing, and/or functionality of the faulty CFTR protein. Every patient diagnosed with CF gets genetic testing to ascertain if their mutation qualifies them for CFTR modulator medication. The selection of treatment is contingent upon the specific mutation and the patient's age. CFTR modulators can be categorized into two groups: potentiators and correctors. Potentiators function by enhancing the transport of chloride ions via the CFTR protein located in the cell membrane. Ivacaftor is a medication that belongs to this class. Correctors are pharmacological agents that influence the spatial conformation of the CFTR protein. Their treatment enhances the intracellular trafficking of CFTR protein and elevates the quantity of its particles in the cell membrane. They ought to be utilized concurrently with ivacaftor. Among the presently available correctors, lumacaftor and tezacaftor are notable. Triple therapy is increasingly significant, involving the concurrent administration of three CFTR modulators: the novel corrector elaxacaftor, in conjunction with tezacaftor and ivacaftor (ETI). In specific patients, CFTR modulator medication enhances pulmonary function, diminishes the frequency of exacerbations, facilitates adequate weight gain, and so elevates the quality of life for individuals with cystic fibrosis. Recent findings from multiple investigations and the outcomes of the extensive SIMPLIFY clinical trial have prompted a shift in treatment strategy, indicating that the broad application of ETI therapy may render ongoing inhalation therapy superfluous for all patients [18][19][20].

In the chronic pharmaceutical management of CF, antibiotics have been identified for both inhalational and oral administration. In patients with chronic *Pseudomonas aeruginosa* infection, chronic inhalation therapy with tobramycin, colistin, or aztreonam lysine is advised; in certain programs, levofloxacin is additionally utilized. For patients with severe lung illness or recurrent exacerbations, particularly those diagnosed with *Pseudomonas aeruginosa* infection, chronic administration of azithromycin is advised due to its anti-inflammatory and immunomodulatory properties [21].

Anti-inflammatory medications may encompass inhaled glucocorticosteroids and oral ibuprofen, albeit only for a specific subset of patients, rather than as standard therapy. Additional inhaled medications that may assist CF patients include bronchodilators; their use is recommended when there is confirmed enhancement in pulmonary function tests or a distinct subjective improvement. Should persistent respiratory failure occur, it may be essential to commence oxygen therapy.

When attending to a patient with CF, it is crucial to prioritize infection prevention. Children afflicted with this condition should receive vaccines comparable to those in the

general population, with particular emphasis on supplementary vaccinations, including pneumococcal vaccination, Covid-19 vaccination, and annual influenza vaccination. Alternative strategies for infection prevention encompass the administration of antiviral medications to specific patients and the implementation of infection control protocols to inhibit pathogen transmission inside healthcare settings.

An additional crucial factor in enhancing the quality of life for children with CF is consistent physical activity. Individuals with CF should be motivated to engage in regular exercise to achieve the same benefits demonstrated for the general population. This aspect of CF management is discussed in the subsequent sections of this work.

4. The role of physical activity in patients with cystic fibrosis

4.1. The impact of physical activity on overall health

The WHO recommends that school-age children engage in a minimum of one hour of moderate to strenuous physical activity daily, incorporating muscle-strengthening exercises 2-3 times per week, whereas preschool children should partake in at least three hours of diverse activities each day. Children diagnosed with CF are not excluded from these recommendations.

Properly selected physical activity reduces the risk of overweight and obesity, hence preventing the development of cardiovascular illnesses, metabolic disorders associated with carbs or lipids, and potentially decreasing the risk of some cancers. Participation in sports positively impacts the psychological development and social integration of children and adolescents. Facilitating the enhancement of coordination, strength, and endurance is equally crucial.

Proper diet for children with CF is a fundamental aspect of managing this condition. Recently, the global epidemic of obesity has exacerbated the issue of excessive body weight among those afflicted with cystic fibrosis [22]. The emergence of novel medicines, including CFTR modulators, presents new chances for individuals with cystic fibrosis to engage in active lifestyles; yet, limitations persist. Some individuals exhibit improper weight gain in response to CFTR modulator therapy, characterized by an increase in fat mass rather than muscle mass, which is a considerable problem for these patients [23]. For overweight young people, consistent physical activity mitigates the risk of cardiovascular and metabolic

disorders, as well as other comorbidities that are more significant in the aging cystic fibrosis population [24].

Physical activity contributes to the overall homeostasis of individuals with CF by reducing insulin resistance, improving bone mass, and supporting proper muscle development. Regular exercise also influences the gut microbiome, which may indirectly modulate immune responses and reduce chronic systemic inflammation.

4.2. The impact of physical activity on the respiratory system

Physical activity plays a crucial role as an adjunct therapy in cystic fibrosis, affecting the respiratory system both mechanically and immunologically. Regular physical exercise enhances mucociliary clearance by increasing ciliary motility in the respiratory epithelium and promoting the secretion of surfactant. This process reduces mucus viscosity and facilitates its removal from the bronchi, thereby decreasing the risk of secretion retention and subsequent.

Moreover, increased pulmonary ventilation during exercise induces bronchodilation and enhances the partial pressure gradient of gases, promoting better tissue oxygenation and reducing secretion retention. A key mechanism involved is the improvement of the ventilation-perfusion (V/Q) ratio, which enables more efficient gas exchange in the alveoli. Simultaneously, the elevated breathing intensity during physical exertion mechanically aids mucus clearance and supports bronchial self-cleaning mechanisms [25].

Regular exercise contributes to an increase in forced vital capacity (FVC) and forced expiratory volume in one second (FEV_1), which are critical parameters for assessing respiratory efficiency. Improvement in these values signifies not only enhanced ventilatory performance but also a deceleration of bronchial obstruction progression, which is a hallmark of disease progression in cystic fibrosis [26].

4.3. Systemic benefits of physical activity – strengthening of respiratory muscles, improved physical fitness and immune system function

Increased activation of the diaphragm and accessory respiratory muscles enhances their strength and endurance, reducing the risk of respiratory fatigue and facilitating the maintenance of effective pulmonary ventilation [27] . Regular exercise improves respiratory mechanics and alleviates dyspnea. Systematic physical activity significantly enhances the

overall fitness of patients with cystic fibrosis, increasing their cardiorespiratory endurance, muscle strength, and general physical performance. Improved physical capacity reduces the subjective sensation of fatigue, allowing patients to perform daily activities with less effort and greater ease. Furthermore, exercise influences respiratory biomechanics by decreasing the work of breathing and improving muscle coordination, which, in the long term, contributes to slowing the progression of obstructive disorders and enhancing the quality of life in patients with CF. Aerobic exercise improves cardiac efficiency by increasing stroke volume and reducing resting heart rate. Enhanced tissue perfusion lowers the risk of cardiovascular complications and supports optimal respiratory function [26].

At the immunological level, regular physical activity exerts a modulatory effect on the immune system, reducing the chronic inflammatory state characteristic of cystic fibrosis [27]. Physical exertion influences the cytokine profile by lowering the levels of pro-inflammatory mediators such as IL-6, TNF-α, and IL-8, which play a key role in neutrophil recruitment to the airways and exacerbation of lung tissue damage. Additionally, exercise induces an increase in anti-inflammatory cytokines, such as IL-10, leading to a reduction in excessive inflammatory responses and oxidative stress. Pulmonary macrophage and regulatory T-cell function is also improved, enhancing their ability to phagocytize pathogens and regulate immune responses. These mechanisms facilitate infection control without exacerbating chronic inflammation.

Regular exercise supports the management of opportunistic infections [28]. Patients with cystic fibrosis are particularly susceptible to chronic bacterial infections, primarily caused by *Pseudomonas aeruginosa*, *Staphylococcus aureus* (including methicillin-resistant MRSA strains), *Burkholderia cepacia complex*, and *Haemophilus influenzae*. The movement of epithelial cilia and dynamic pressure changes in the bronchi during exercise promote the clearance of bacterial biofilm, which is highly resistant to antibiotic therapy and serves as a reservoir for persistent infections [25]. Among fungal infections, *Aspergillus fumigatus* is a common pathogen in individuals with cystic fibrosis; breathing exercises and improved tissue oxygenation can also help mitigate conditions favorable for fungal colonization [26].

Moreover, physical activity may reduce the risk of viral infections, including those caused by *Rhinovirus*, *Influenza virus*, and *Respiratory Syncytial Virus (RSV)*, which frequently lead to disease exacerbations and secondary bacterial infections in cystic fibrosis patients. This

mechanism involves an increased production of type I and III interferons, which play a crucial role in inhibiting viral replication and enhancing the expression of genes associated with innate immunity.

4.4. The Role of Exercise in Preventing Pulmonary Complications

Physical exertion also influences the body's redox balance by reducing reactive oxygen species (ROS) levels in the lungs through the activation of antioxidant enzymes such as superoxide dismutase (SOD) and catalase (CAT) [27] . This limits lipid peroxidation in bronchial epithelial cell membranes and protects against oxidative damage, a key mechanism driving the progression of obstructive pulmonary changes. In the context of preventing pulmonary complications, physical activity not only supports bronchial drainage but also plays a crucial role in reducing opportunistic infections. In the long term, this translates into a slower progression of obstructive disorders and an improved quality of life for patients with CF [25]. The incorporation of systematic training into standard patient care has been shown to extend survival.

Certain CF patients have choose to utilize physical activity as an alternative to traditional airway clearing techniques; nevertheless, the long-term efficacy of exercise as an airway clearance therapy necessitates additional research, while its short-term effects are presently under investigation in the ExACT clinical trial [29] [30].

The benefits of appropriately planned regular physical activity in patients with cystic fibrosis are summarized in Table 1.

Benefits of physical activity in CF patients	
general health condition	respiratory system
• maintaining a healthy body weight • improving physical fitness • modulating effect on the immune system, reducing chronic inflammation • reducing insulin resistance • improving bone mass • beneficial effect on the intestinal microbiome	• enhancing mucociliary clearance, supporting bronchial self-cleaning mechanisms • bronchodilation and improvement of the ventilation-perfusion (V/Q) ratio • increasing in FVC and FEV_1, deceleration of bronchial obstruction progression, improvement of respiratory capacity • strengthening of respiratory muscles • protection against oxidative damage to the respiratory epithelium

Table 1. The benefits of physical activity in CF patients

5. Physical Activity in Children with Cystic Fibrosis – Age-Appropriate Adaptation and Exercise Types

Physical activity plays a crucial role in the treatment of children with cystic fibrosis, contributing to improved pulmonary function, airway clearance mechanisms, and overall physical endurance. Regular physical exertion supports bronchial clearance by increasing airflow, mobilizing mucus, and strengthening respiratory muscles [25][31]. Current guidelines from the European Cystic Fibrosis Society (ECFS) and the American Thoracic Society (ATS) recommend incorporating systematic physical activity as an integral part of CF management, tailored to the patient's age and disease severity [32]. Recommended types of exercise include aerobic, resistance, and breathing exercises, as well as movement-based play, which is particularly beneficial for young children as a complementary physiotherapy approach.

In infants, the primary goal of physical activity is to support airway clearance mechanisms and stimulate proper chest mobility. At this stage, passive physiotherapy techniques, such as postural drainage and chest vibrations, are commonly employed to facilitate mucus transport

toward larger bronchi, where it can be effectively removed [25]. Postural drainage involves positioning the infant to enhance mucus transport to the upper airways, a standard practice in newborns and infants [32]. Additionally, techniques promoting diaphragmatic breathing and early motor activities, such as crawling and creeping, help strengthen respiratory muscles and improve lung ventilation [25]. Some studies suggest potential benefits of using positive expiratory pressure (PEP) therapy in this age group, though its efficacy remains inconclusive and requires further research [25] [33].

In preschool-aged children, physical activity should include both breathing exercises and dynamic movement-based activities that support proper lung ventilation. Active Cycle of Breathing Techniques (ACBT) is particularly effective in this age group. This method consists of three phases: breath control, deep breathing, and forced expiratory technique (huffing) [32]. Huffing, taught through playful activities, facilitates mucus clearance and enhances breathing mechanics. Physical activities should be engaging and enjoyable for the child, with recommended exercises including bubble blowing and balloon inflation, which actively engage respiratory muscles.

For children in early school years, physical activity should be regular and of increased intensity. Studies show that aerobic exercises such as running, cycling, and swimming significantly improve spirometric parameters, including forced expiratory volume in one second (FEV_1), and enhance exercise tolerance[32]. Swimming is particularly beneficial as it engages all muscle groups while controlled breathing improves coordination and respiratory muscle efficiency. At this stage, strength training is also introduced to enhance muscle strength and postural stability. Resistance exercises, such as bodyweight training (e.g., squats, push-ups), reduce the risk of chest wall deformities and improve pulmonary ventilation efficiency [25]. Additionally, children in this age group may use airway clearance devices such as PEP and oscillatory PEP (OPEP) devices, which increase airflow through the bronchi and facilitate mucus clearance [32].

In adolescents, physical activity should focus on maintaining respiratory fitness and slowing lung function decline. Interval training, which alternates short bursts of intense activity with rest periods, is one of the most effective methods for improving physical endurance in this age group. Additionally, resistance training is recommended to support muscle development and chest stabilization, preventing osteopenia, a common complication

in CF patients [25] . Autogenic drainage (AD) is an airway clearance technique that involves controlled regulation of breath volume. It consists of three phases: shallow breaths to mobilize mucus, medium-depth breaths to move secretions to larger bronchi, and deep breaths with controlled exhalation to facilitate mucus removal. AD has been shown to be more effective than traditional airway clearance methods, reducing the need for forceful coughing and minimizing respiratory muscle fatigue [32].

6. Safety and recommendations for parents and guardians

Physical activity plays a key role in the treatment of cystic fibrosis, supporting respiratory function, improving fitness and overall quality of life. However, it should be tailored to the individual child's abilities and health status to avoid overstressing the body.

Physical activity is playing an increasingly important role in the rehabilitation of patients with cystic fibrosis. It is an effective and easy-to-learn method that does not require a lot of money. Regular exercise has become a standard part of therapy, and children should be encouraged to engage in various forms of movement from an early age. A key role is played by parents, who can shape positive attitudes toward physical activity in their children through their own attitudes and habits. In the United States, it is recommended that school-aged children spend at least an hour a day in moderate physical activity. Unfortunately, low levels of physical activity are often observed among children with cystic fibrosis due to, among other things, excessive parental concern, fear on the part of the children themselves, or the belief that intense exercise can harm them [34].

A key factor affecting the effectiveness of therapy is its regularity and starting as early as possible. With the introduction of newborn screening, it has become possible to make a quick diagnosis and thus implement appropriate treatment early. Physiotherapists play an important role not only in supporting parents, but most importantly in educating them. They should learn about available physiotherapy methods and forms of physical activity suitable for their children. To increase their awareness, it is essential to present the benefits of regular exercise, which can significantly improve the patient's health. Systematic physical activity contributes to increased chest mobility, improved lung ventilation and increased exercise tolerance. In addition, lung pressure increases during exercise, which facilitates the delivery of air to the small bronchi, thereby supporting the functioning of the respiratory system [34][35].

In order to provide the patient with an effective and, above all, safe exercise program, regular monitoring is necessary. During physical activity, the patient's condition should be

carefully observed, with particular attention to the breathing rhythm, which should be calm and even. It is recommended that inhalation be through the nose and exhalation through a slightly tightened mouth, which promotes the generation of so-called positive expiratory pressure. Such breathing techniques improve breathing efficiency and prevent bronchial collapse.

Suggested types of physical activity include:

- Cardiovascular exercises – activities like swimming, cycling, moderate-paced running, walking, and dancing.
- Muscle-strengthening workouts – light resistance training and exercises using body weight.
- Respiratory exercises – techniques designed to aid mucus clearance, such as the Active Cycle of Breathing Technique.

7. Strategies to support and maintain an active lifestyle

A study published in 2024 [36] indicates diminished physical activity levels and functional performance among youth patients with CF, highlighting a critical necessity for programs that promote physical activity and sports within this patient population. Consequently, the encouragement of an active lifestyle in CF patients should commence in early childhood, significantly impacting the patient's immediate family. During preschool and school years, children ought to be encouraged to engage in diverse organized and club activities, while the specialized team responsible for the patient should oversee the assessment of exercise ability and advancements in this domain. Exercise regimens must be tailored to individual factors such as age, gender, disease severity, presence of comorbidities, and patient preferences, which are crucial for sustaining long-term physical exercise. Patients and their guardians should get competent instruction regarding the safety and advantages of physical activity. Identifying potential physical and psychosocial impediments is essential for effective counteraction [37].

Due to technology advancements, doctors can now provide patients with a range of electronic gadgets for measuring physical activity [38]. They assist patients in attaining daily activity objectives, serve as effective instruments for self-regulation, and enhance motivation. The integration of physical activity with virtual reality games represents an innovative strategy that has demonstrated an enhancement in physical activity levels among young patients suffering from diverse chronic conditions, including hemophilia, cerebral palsy, and

post-severe burn recovery. Investigation is required about the efficacy of this type of physical activity for children and adolescents with cystic fibrosis, as young patients find such technological innovations particularly appealing and adapt effectively to their utilization [39] [40] [41].

8. Summary

Cystic fibrosis is a congenital genetic disease caused by a mutation in the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene. The CFTR gene is responsible for producing a protein that regulates chloride ion transport across cell membranes, leading to excessive mucus thickening in the body. This results in chronic lung infections, pancreatic insufficiency, liver disease, and reduced fertility. CF is most common in Europeans and affects many systems, particularly the respiratory and digestive systems, leading to metabolic problems and difficulties with nutrient absorption.

Respiratory symptoms of CF typically begin in early childhood with a recurring cough that evolves into a chronic condition. Disease progression is marked by abrupt exacerbations, heightened cough, accelerated respiration, dyspnea, and augmented sputum output. Chronic sinusitis is also common, causing structural damage and symptomatic polyps.

Cystic fibrosis requires interdisciplinary management, including pulmonologists, physiotherapists, dietitians, psychologists, and other specialists. The primary aspect of chronic treatment is clearing retained secretions from the respiratory tract, ensuring adequate nutrition, preventing and effectively managing bacterial infections, mitigating inflammation, and preventing complications. Lung transplantation is a treatment option for severely advanced lung disease.

Physical activity is a vital adjunct therapy in cystic fibrosis, affecting the respiratory system both mechanically and immunologically. Regular exercise enhances mucociliary clearance, reducing mucus viscosity and facilitating its removal from the bronchi, thereby decreasing the risk of secretion retention and subsequent pulmonary infections. Increased pulmonary ventilation during exercise induces bronchodilation and enhances the partial pressure gradient of gases, promoting better tissue oxygenation and reducing secretion retention. Elevated breathing intensity during physical exertion mechanically aids mucus clearance and supports bronchial self-cleaning mechanisms.

Physical activity also contributes to systemic benefits, such as strengthening respiratory muscles, improving physical fitness, and modulating the immune system. It reduces the

chronic inflammatory state characteristic of cystic fibrosis, lowering pro-inflammatory mediators and increasing anti-inflammatory cytokines. Exercise supports the management of opportunistic infections, particularly bacterial infections, and may reduce the risk of viral infections. Furthermore, physical activity contributes to overall homeostasis by reducing insulin resistance, improving bone mass, and supporting proper muscle development.

Physical activity is crucial in the treatment of children with cystic fibrosis, contributing to improved pulmonary function, airway clearance mechanisms, and overall physical endurance. Guidelines from the European Cystic Fibrosis Society (ECFS) and the American Thoracic Society (ATS) recommend incorporating systematic physical activity as an integral part of CF management, tailored to the patient's age and disease severity. Recommended types of exercise include aerobic, resistance, breathing exercises, and movement-based play.

Physical activity is an effective, easy-to-learn method that can be tailored to individual children's abilities and health status. Parents play a key role in shaping positive attitudes towards physical activity in their children. Regular monitoring is necessary to ensure a safe exercise program. Physiotherapists play a vital role in educating parents about available physiotherapy methods and forms of physical activity suitable for their children.

Disclosure

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