Fisioterapia Respiratória em Pacientes com Fibrose Cística

Respiratory Physiotherapy in Patients with Cystic Fibrosis

Fisioterapia Respiratoria en Pacientes con Fibrosis Quística

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REVISA

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RESUMO

Objetivo: O objetivo deste estudo foi proporcionar uma visão sobre a FC e as possíveis formas de tratamento fisioterapêutico. **Método:** Trata-se de uma revisão de literatura narrativa. Foram usadas as bases de dados eletrônicas: LILACS, SCIELO, PUBMED e MEDLINE a partir do ano de 2008. Na coleta de dados foram usados os seguintes descritores: mucoviscidose, fibrose cística, métodos de avaliação e fisioterapia respiratória. Foram selecionados artigos relacionados a avaliações e tratamento da FC, publicados em português e inglês. **Resultados:** A fisioterapia respiratória é recomendada para os pacientes com FC por possuir técnicas efetivas para aumento da expectoração de secreção, no entanto ainda se faz necessária a comprovação científica, em longo prazo, dos efeitos de cada técnica. **Conclusão:** Os tratamentos mais usados e atuais de acordo com os artigos encontrados foram drenagem postural, vibrocompressão, drenagem autogênica, aceleração do fluxo expiratório, expiração lenta total com a glote aberta em decúbito infralateral, máscara de pressão positiva expiratória e exercícios aeróbicos. Sendo elas importantes para amenizar os sinais respiratórios que a mucoviscidose apresenta evitando possíveis complicações pulmonares.

Descritores: Mucoviscidose; Fibrose Cística; Fisioterapia Respiratória; Métodos de Avaliação.

ABSTRACT

Objective: The objective of this study was to provide an insight into CF and the possible forms of physiotherapeutic treatment. **Method:** It is a narrative review of literature. The electronic databases were used: LILACS, SCIELO, PUBMED and MEDLINE from 2008. In data collection, the following descriptors were used: mucoviscidosis, cystic fibrosis, assessment methods and respiratory physiotherapy. Articles related to CF assessments and treatment, published in Portuguese and English, were selected. **Results:** Respiratory physiotherapy is recommended for patients with FC for having effective techniques for increasing sputum secretion, however it is still necessary to prove long-term scientific analysis of the effects of each technique. **Conclusion:** The most used and current treatments according to the articles found were postural drainage, vibrocompression, autogenic drainage, acceleration of expiratory flow, total slow expiration with the open glottis in the infralateral decubitus, positive expiratory pressure mask and aerobic exercises. They are important to mitigate the respiratory signs that mucoviscidosis presents, avoiding possible pulmonary complications..

Descriptors: Mucoviscidosis; Cystic fibrosis; Respiratory fisioterapy; Assessment Methods.

RESUMEN

Objetivo: El objetivo de este estudio fue proporcionar una idea de la FQ y las posibles formas de tratamiento fisioterapéutico. **Método**: Esta es una revisión de la literatura narrativa. Se utilizaron las bases de datos electrónicas: LILACS, SCIELO, PUBMED y MEDLINE de 2008. En la recopilación de datos, se utilizaron los siguientes descriptores: mucoviscidosis, fibrosis quística, métodos de evaluación y fisioterapia respiratoria. Se seleccionaron los artículos relacionados con las evaluaciones y el tratamiento de la FQ, publicados en portugués e inglés. **Resultados:** Se recomienda fisioterapia respiratoria para pacientes con FC por tener técnicas efectivas para aumentar el esputo secreción, sin embargo, todavía es necesario demostrar Análisis científico a largo plazo de los efectos de cada técnica. **Conclusión:** Los tratamientos más utilizados y actuales según los artículos encontrados fueron drenaje postural, vibrocompresión, drenaje autógeno, aceleración del flujo espiratorio, espiración lenta total con glotis abierta en posición de decúbito infralateral, máscara de presión espiratoria positiva y ejercicios aeróbicos. Son importantes para mitigar los signos respiratorios que presenta la mucoviscidosis, evitando posibles complicaciones pulmonares.

Descriptores: Mucoviscidosis; Fibrosis quística; Fisioterapia respiratoria; Métodos de evaluación.

REVIEW

Introduction

Cystic fibrosis (CF) is also known as mucoviscidosis, being a genetic disease of an autosomal recessive character, characterized by an imbalance of ionic transport, which causes an increase in the viscosity of secretions, causing obstruction of the gland ducts, inflammation, injury and destruction progressive tissue, with multisystemic consequences.[1] The disease in whites is quite common. The Brazilian Registry of Cystic Fibrosis (REBRAFC) reports that survival is around 43.8 years analyzed in the year 2016. The data points to approximately 4,654 records of people diagnosed with cystic fibrosis (CF).[2] Rio Grande do Sul is the second state with the largest number of patients, covering 356 individuals (13.3%). The pediatric population (under 18 years old) is 1,918 individuals (77.6% of patients in the state).[3-4]

The functioning of the protein called Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) is compromised. This protein is responsible for maintaining the normal flow of ions in the epithelial cells of the respiratory system, pancreas, liver and reproductive system, its dysfunction will cause a decrease in the permeability of cells to chloride causing changes in chloride secretion in the cells and an increase in sodium reabsorption. , favoring that the secretions are thick. The exocrine glands involvement can lead to malnutrition, coughing, infection of bacteria in the airways being predominant.[5-6]

The most common respiratory signs of mucoviscidosis are: persistent chronic cough and excessive secretion production. Which can be perceived through the stethoscope mainly lung sounds called wheezing snores.[7] It is a pathology that, despite affecting many organs, the respiratory tract is considered the most harmful, being responsible for the highest number of deaths.[8-9] According to Mauch, Kmit and Marson (2016) and Silva and Lima (2018) the changes generated in the respiratory system by the pathophysiology of cystic fibrosis are:

Changes in ionic transport that lead to increased viscosity of secretions, making them thicker and leading to a chronic inflammatory process. Inflammation can lead to various impairments, in the most severe cases respiratory failure and death. Changes if left untreated can lead to pneumothorax, hemoptysis, pneumonias, atelectasis, bronchiectasis, bronchitis, cor pulmonale and respiratory failure.[10-11]

In view of the variety and chronicity of the affections, each patient undergoes an individual evaluation, where a specific treatment must be elaborated, aiming at their greatest deficiencies, thus, there is no standard model for the treatment of CF. The earlier and performed by a multidisciplinary team, the greater the chances of delaying the progression of the pathology, as well as offering a better quality of life.[38,35] As a result, the survival of these patients tends to increase. Arriving to adulthood, new complications may arise, often requiring surgical procedures.

In this context, the objective of this study was to provide an insight into cystic fibrosis and describe the best forms of treatments for a better prognosis for these patients.

Method

This is a review of narrative literature. From journals indexed in the reference portals found in the LILACS, SCIELO, PUBMED and MEDLINE databases.

In data collection, the following descriptors were used: mucoviscidosis, cystic fibrosis, assessment methods and respiratory physiotherapy. The research of the work was based on the inclusion and exclusion criteria. The inclusion criteria favored publications starting in 2008, using languages in Portuguese and English. The works were selected by reading the title, abstracts and keywords. In the exclusion group, studies that did not present a coherent subject regarding the study objective were discarded.

Assessment methods for patients with cystic fibrosis

Pulmonary auscultation

Pulmonary auscultation is a simple and low-cost method of physical assessment of the chest, which aims to identify and assess lung sounds through the stethoscope, these can be normal or pathological. The lung sounds considered normal are the physiological noises attributed to the breath sounds, while the pathological sounds also known as adventitious noises are the crackles, snores, crackles and wheezing. [12]

It needs to be carried out in a quiet and calm place, with the chest bare, the patient is in an orthostatic position, erect, the therapist starts by listening to the anterior part, then after listening to the posterior part, the patient must be instructed to take a deep breath through his mouth thus having a better identification of the sounds present. A good interpretation must be made as to the type, location, and intensity of the breathing sounds present.[13]

Medical Scale Research Council (MRC)

MRC is used to measure the degree of difficulty in breathing during daily activities, where a table is given to the patient where he will have to graduate the degree of dyspnea from 1 to 5 as shown in Chart 1.

Quadro 1. *Graduação da escala MRC*

Escala	Características
1	Tem falta de ar ao realizar exercício intenso.
2	Tem falta de ar quando apressa o passo ou sobe ladeiras ou escadas.
3	Precisa parar para respirar mesmo andando devagar, ou anda lentamente em comparação com pessoas da mesma idade.
4	Necessita parar muitas vezes devido a falta de ar quando anda cerca de 100 metros, ou poucos minutos de caminhada.
5	Sente tanta falta de ar, que não sai de casa e precisa de ajuda para realizar atividades de vida diária (AVDS).

Fonte: adaptado de Demetria et al. (2008)

Bioimpedance (BIA)

Method used to measure body composition indicating weight (kg), percentage of water, bone mass, fat and lean mass, through the passage of low-intensity electrical current in biological tissues.[15]

In order to avoid interference during this method, it is necessary to follow some guidelines.[16]

Peak Expiratory Flow (PEF)

To perform the PEF test, the patient must remain seated in an armless chair, and instructed to keep the mouth completely attached to the mouthpiece, to avoid air leakage during the test. The therapist should advise the patient to take a maximum inspiration, followed by a voluntary cough (rapid, short and explosive exhalation), performing a minimum of three repetitions. If there was a difference between the first three attempts greater than 10% or 20 l / min, allow the test to be performed in up to 10 attempts, using the highest value as a result.[17-18] The main objective of this test is to assess the peak expiratory flow, thus being able to quantify the degree of airway obstruction.

Treatment methods for patients with cystic fibrosis

Bronchial hygiene techniques

Respiratory physiotherapy is used to treat patients with cystic fibrosis in order to minimize the pulmonary manifestations that the pathology presents. The most used techniques according to the literature are: Postural drainage, tapping, vibrocompression, autogenic drainage, Expiratory Flow Acceleration (AFE), Total slow exhalation with the open glottis in the infralateral decubitus position. (ELTGOL) and expiratory positive pressure maneuver [17].

Postural Drainage

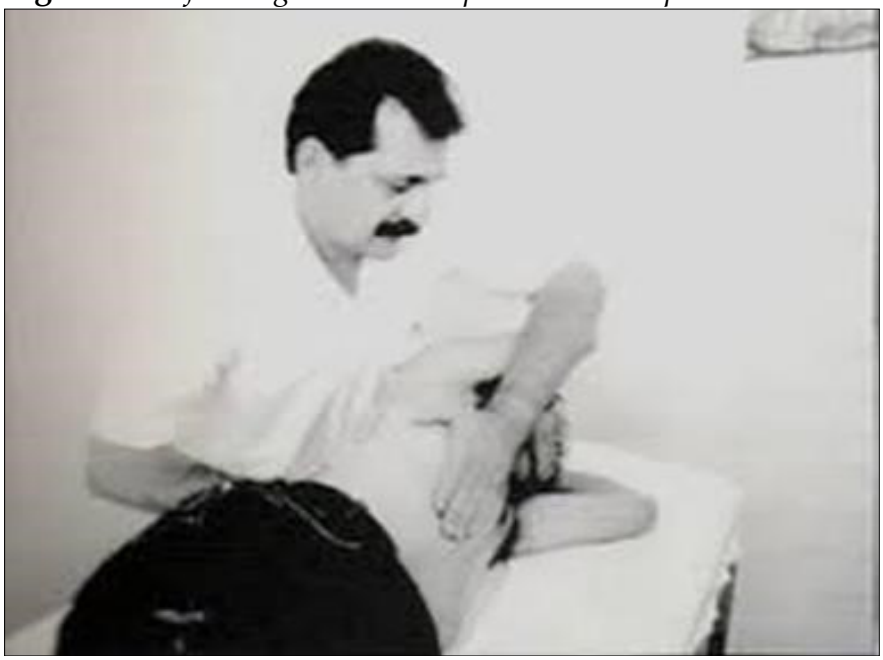
Postural drainage uses the gravitational effect, employing the inverted position, for example, patient in decubitus with the hip higher than the shoulders. Drainage is performed by segments of the lung, there are twelve positions that can be used, among which at least nine must be used in order to have complete drainage in all areas of the lungs and must be maintained for three to fifteen minutes each in one frequency three to four times a day. It aims to drain the pulmonary secretion of the bronchial tree using the action of gravity, being widely used together with other techniques in order to enhance its action.19 To avoid complications during this technique, the therapist should also be aware of the contraindications.[20]

Vibrocompression

In this technique the hands are placed directly on the skin on the chest wall and a slight compression is performed, and at the same time, a vibration on the chest wall, in the same direction as the chest is moving. This vibratory action

occurs through the isometric contraction of the therapist's upper limb, whose main objective is to use the patient's position to facilitate the drainage of pulmonary secretions towards the trachea so that it is easily expectorated or aspirated. Its indications are for patients with hypersecretion, on mechanical ventilation, smokers and neuromuscular disorders, and the contraindications are in case of subcutaneous emphysema when there is pain, osteoporosis, rib fracture and obese patients 21, according to Figure 1.

Figure 1- *Performing the vibrocompression technique*



Source: Castro et al. (2010)

Autogenic Drainage

In this technique, the patient will be seated, erect and will take a deep breath at a normal or relatively slow pace, with this the secretions in the airways will move into the trachea as a result of the breathing pattern, and as the secretions move to the trachea they will be expelled with a soft cough or a slightly forced exhalation. It has the objective of increasing the air flow in the airways so that it improves the elimination of mucus and thus also improves pulmonary ventilation. This technique is used as an indication for patients who cooperate and do not need manual assistance from the therapist, being contraindicated in a patient who has intolerance to handling the technique.[22]

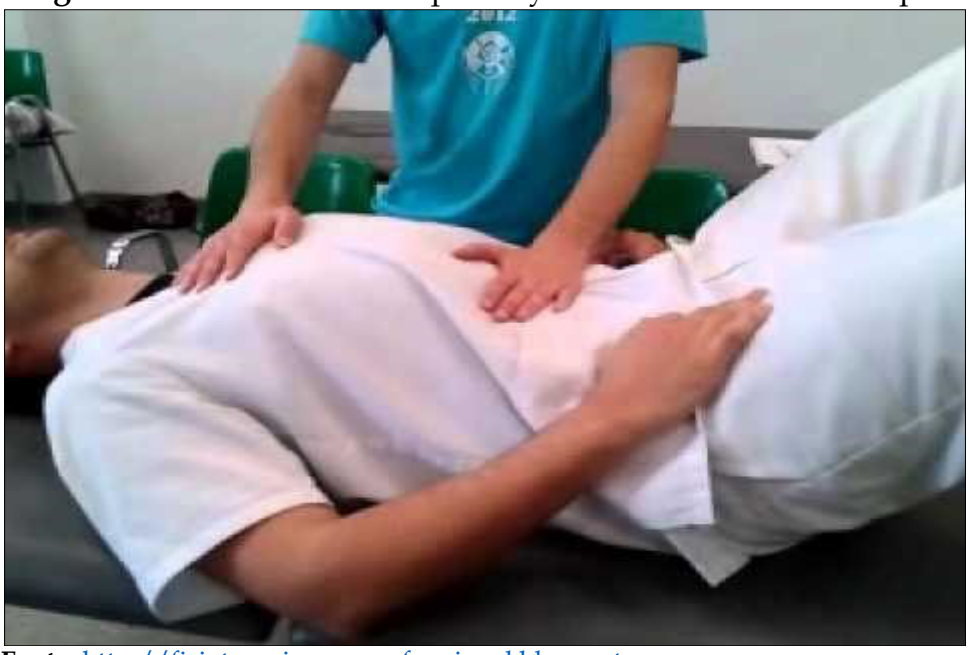
Acceleration of Expiratory Flow (AFE)

The expiratory flow acceleration technique can be performed with the patient in the lateral decubitus or supine position. In supine, the therapist places his hands on the patient's parasternal regions, following the chest movement in the expiratory phase, applying pressure at the end of expiration, thus prolonging the expiratory phase even more, this pressure is applied in the craniocaudal and lateral lateral directions. In lateral decubitus the pressure is applied in the same direction and with only one hand, while with the other hand, a friction in the

posterior thoracic region should be performed, helping compression and protecting the costovertebral joints.[23]

The technique aims to deflate the chest and lungs, thereby reducing the residual space and increasing the volume of tidal air, increasing pulmonary ventilation and, consequently, better oxygenating the blood. It also aims to use the energy applied by the physiotherapist on the patient's chest with the intention of improving the mobility of the rib cage and may, in its final stage, stimulate coughing, inciting the elimination of secretion. It is contraindicated in the acute initial phase of bronchiolitis and asthmatic crisis, which is still not very secretive[24], as shown in Figure 2.

Figure 2- Execution of the expiratory flow acceleration technique

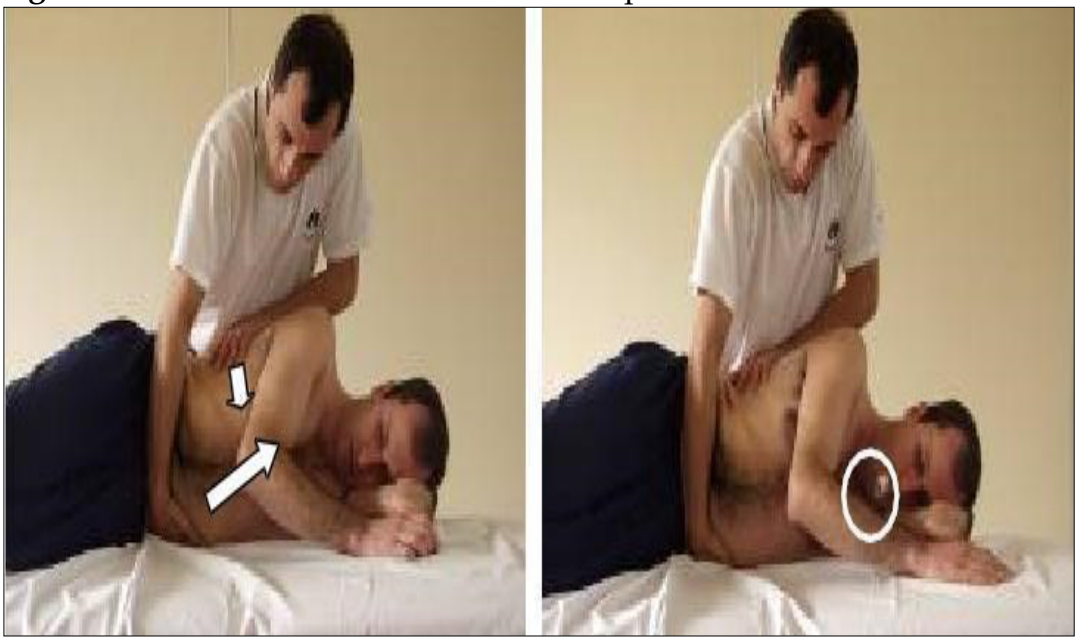


Fonte: http://fisioterapiapneumofuncional.blogspot.com

Total Slow Expiration with Open Glottis in Infralateral Decubitus (ELTGOL)

Before starting the technique, pulmonary auscultation is performed to locate the region where the secretion is accumulated. After this procedure, the patient is positioned in lateral decubitus with the lung compromised in an infralateral position, which contributes to the removal of secretion due to the greater diagrammatic path and greater disinflation, which increases expiratory time and expiratory flow. The physiotherapist positions himself behind the patient and with one hand will exert infralateral abdominal pressure, which favors the emptying of the affected lung volume, and with the other hand, he will perform a counter-support pressure at the costal level. While the physiotherapist performs abdominal pressure, the patient is instructed to breathe the tidal volume and exhale with the open glottis to the residual volume, starting from the residual capacity. With the general objective of the technique, that of using lateral positioning to facilitate the elimination of mucus 25, according to Figure 3.

Figure 3- Realization of the ELTGOL technique.



Source: Guimarães, et al. (2012)

Positive expiratory pressure mask (PEP)

The use of PEP in respiratory physiotherapy was described in 1984. The system consists of a face mask attached to a unidirectional valve, in which a resistance of 10 to 20 cmH2O is determined in the expiratory phase. The patient inhales and exhales inside the mask for approximately 15 times or 2 minutes. After resting with breaths out of the mask, the procedure is repeated for approximately 20 minutes and usually performed twice a day. Increased secretion sputum, improved lung function and oxygen saturation have been demonstrated when comparing PEP to PD, percussion and breathing exercises in 14 fibrocystic patients. In addition, the effects of the PEP mask with DP associated with percussion were compared in a group of 40 CF patients for a period of 1 year and observed an increase in lung function, as assessed by FVC, FEV1 and FEF25-75 %, in patients who used PEP. PEP is also the most indicated resource than PD for fibrocystic patients with reflux symptoms. In short-term studies, the PEP mask was effective in the mobilization of secretions and, in the long term, in the maintenance of pulmonary function in fibrocystic patients.[26] However, in another study, fibrocystic patients who performed breathing exercises, TEF and DP expectorated significantly more amounts of secretion than those who used the PEP mask in the treatment. In a Cochrane review, 20 randomized studies were selected that compared the effectiveness of PEP with conventional methods of physical therapy in fibrocystic patients. The authors concluded that there is no clear evidence that PEP is more or less effective than other methods in bronchial hygiene and in improving lung function in these patients.[27]

Principles of physical training

As described above, patients with cystic fibrosis have malnutrition, peripheral and respiratory muscle atrophy. To improve these signs, physical training must respect four general principles of physiology: overload, reversibility, intensity and specificity.[28]

The overload principle refers to the observation that a system or tissue must be exercised at a level beyond which it is used for the training effect to occur, the system or tissue gradually adapts to this overload. The reversibility principle, on the other hand, only indicates that gains are quickly lost when the overhead is removed. The third principle is the intensity that constitutes the frequency and duration of the exercise. The fourth is the principle of specificity where training is specific to the muscle fibers involved in the activity, in addition the muscle fibers adapt specifically to the type of activity, mitochondrial and capillary adaptations to resistance training and adaptations of contractile proteins to strength training with Weight.[29] For this principle, we can take aerobic exercises as an example.

Aerobic exercises

Selvadurai and Meyers (2002) analyzed the interference of the practice of aerobic exercise in the maximum oxygen consumption (VO2 max) during the exercise test. The improvements in exercise tolerance during aerobic training were significant when compared to the group that did not perform any specific training. Periods of desaturation were observed in the aerobic training group, but there was no significant difference between the groups in the increase in FEV.[1] In this same study, the aerobic training group showed a significant increase in the strength of the lower limbs when compared to the control group (without physical activity). In addition, the authors assessed patients' quality of life using the Quality of Well-being Scale and demonstrated a significant difference between groups in changing quality of life.

Results and Discussion

Respiratory physiotherapy is recommended for CF patients because it has effective techniques for increasing secretion sputum, however, long-term scientific proof of the effects of each technique is still necessary.

When selecting a bronchial hygiene therapy, physiotherapists should consider some fundamental factors, such as motivation and goals of the patient, ability to concentrate and ease of learning, effectiveness of the technique, necessary respiratory work, need for assistants or equipment, costs, advantages of the combination of the patient's methods and age. Cost is a factor that is becoming critical in the selection of treatment strategies.

Of course, considering everything else equal, we should always select the least expensive strategy. Although it has been elucidated in the literature that the performance of respiratory physiotherapy in fibrocystic patients is essential, there is still no consensus regarding the most effective form of intervention.

The use of conventional methods such as Postural Drainage and Tapotage exclusively was not validated in any work. Although questioned and

abolished by many, this technique needs investigation not only as to the effect but as to the form of realization, respecting the use of both hands simultaneously and the number of percussions per minute.

As for Percussion and Vibrocompression performed mechanically, although they do not replace an experienced physiotherapist, they do not cause muscle fatigue and can provide constant frequencies, rhythms and impact forces. However, there is no strong evidence that these devices are more effective than performing the techniques manually.

The Technique of acceleration to expiratory flow has good results when it comes to mobilization of secretion, but with contradictory effects in improving lung function. Regarding the PEP mask, there is no clear indication that this resource is more or less effective than the others. It seems that its effect is more related to improvements in lung function than especially in bronchial hygiene.

Autogenous drainage shows good results in relation to mobilization of secretion and improvement of lung function, with the advantage of being less likely to produce oxygen desaturation. However, it has the disadvantage of needing a long session and technique guidance regularly.

In addition to these techniques, studies by Habib et al., 2015 have also shown the interference of regular exercises in the lives of patients with CF. However, there is still no consensus on the ideal training program. What is clear is that regularity and adherence to the exercise program are essential. The support of family members is very important for the continuity of the exercises proposed by the service team. Careful assessments of physical fitness and clinical status show that most patients with CF are able to practice regular physical activity. It is up to the professionals involved in the care to provide information to patients and their families, in order to ensure the adoption of adequate habits of physical exercise for maintenance and improvement of health.

Respiratory physiotherapy is synonymous with better quality of life for fibrocystic patients and one of the most important treatment tools. However, it is essential to carry out more studies, especially in the long term, with the performance of maneuvers in a more standardized way to really point out the most effective physiotherapeutic resources in the treatment of these patients. However, it is important to consider that motivation is the key to adherence to any procedure, especially for chronic outpatients or those treated at home, as none method will be successful if it is abandoned by the patient.

Conclusion

Which respiratory symptoms, such as impaired lung function and respiratory exacerbations, were the factors that most affected the quality of life of adolescents and adults with CF, a role of the physiotherapist that becomes essential with multidisciplinary care with regard to vision acceptable value of the patient in relation to the disease.

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