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Cystic fibrosis - Ways to improve the quality of life of patients

Kamila Świercz
Wojewódzki Szpital im. Św. Ojca Pio w Przemyślu
https://orcid.org/0000-0003-0783-270X

Magdalena Majcher
1 Wojskowy Szpital Kliniczny w Lublinie
https://orcid.org/0000-0003-3998-090X

Monika Majcher
Wojewódzki Szpital Specjalistyczny im. Stefana Kardynała Wyszyńskiego w Lublinie
https://orcid.org/0000-0002-4924-9048

Dominika Górska
1 Wojskowy Szpital Kliniczny z Polikliniką w Lublinie
https://orcid.org/0000-0002-2444-4891

Aleksandra Karwańska
5 Wojskowy Szpital Kliniczny z Polikliniką w Krakowie
https://orcid.org/0000-0001-6917-6985

Agata Pikulicka
SZPITAL SOLEC w Warszawie
https://orcid.org/0000-0003-1693-8127

Małgorzata Sierpień
1 Wojskowy Szpital Kliniczny z Polikliniką SPZOZ w Lublinie
https://orcid.org/0000-0002-0119-2775

Piotr Brzychczy
Szpital im J.Dietla w Krakowie
https://orcid.org/0000-0002-9607-9942

Aleksandra Kulbat
V Wojskowy Szpital Kliniczny z Polikliniką w Krakowie
https://orcid.org/0000-0002-3981-4242

Mateusz Kulbat
V Wojskowy Szpital Kliniczny z Polikliniką w Krakowie
https://orcid.org/0000-0003-3421-5512

Abstract

Introduction

Cystic fibrosis (CF) is a serious, multi-system disease. In the Caucasian population CF is the most common autosomal recessive disease, which happens for 1 person in 3,500 births [1]. In the course of CF, there is a dysfunction of many organs, mainly the respiratory and digestive systems. Symptomatic treatment of cystic fibrosis is very important. The change in daily eating habits, the use of proper breathing techniques and psychotherapy also have an impact on improving the quality of life of patients.

In the past patients died at a young age. The progress in the diagnosis and treatment of cystic fibrosis has meant that nowadays more and more people are reaching adulthood. In recent years, the median survival of patients has increased to 46.2 years. [2]

Purpose

The aim of this review is to present the current state of knowledge about ways to improve the quality and length of life of patients with CF.

Methods

The literature available in the Pubmed and Google Scholarship databases was reviewed using key phrases.

Results

Additional methods of treatment, such as: learning the correct breathing technique, oxygen therapy, physical exercise or proper nutrition, significantly improve the living conditions of patients.

Keywords: cystic fibrosis; treatment; health education; life comfort; quality of life;

Introduction

Cystic fibrosis (CF) is a congenital genetic disease. Since 1989, it is known that it is caused by a mutation in the CFTR gene (transmembrane conductance regulator of cystic fibrosis) [2]. The gene is located on the long arm of chromosome 7 at locus 7q31.2. In more than 90% of patients with cystic fibrosis, it is possible to establish the mutation leading to the disease. Over 2,000 different mutations of the CFTR gene have been detected, the most common being F508del. The F508del mutation is present in 82.4% of Europe cystic fibrosis patients. Statistics show that more patients are in Northern Europe than in Southern Europe with this mutation. [4] Depending on the type of mutation, the symptoms of the disease may vary. From only elevated sweat chloride values to multi-organ dysfunction with fatal outcome.

However, the stage of the disease does not depend only on the genetic mutation. They are also influenced by factors such as genetic predisposition, lifestyle or individual reactions of the organism. Individual patients may respond differently to identical treatments. [5]

A mutation of the chloride channel protein in the plasma membrane disrupts secretion by the exocrine glands. Thick, residual mucus causes organs malfunction such as the lungs, pancreas, intestines, bile ducts and vas deferens. This results in numerous respiratory infections, pancreatic exocrine insufficiency, malabsorption syndromes, diabetes and infertility.

Airway clearance techniques

Airway Clearance Techniques (ACTs) are activities aimed at increasing the clearance of mucus in the respiratory system, thereby reducing the number of infections and bothersome symptoms such as coughing and shortness of breath. Eliminating these ailments is very important as respiratory failure is the leading cause of death in cystic fibrosis patients. [6] In the past, the only 'gold standard' was manual patting of the chest, which is still used today in children and infants. Currently, ACTs include techniques such as postural drainage and percussion (PD&P), active breathing cycle techniques (ACBT), autogenous drainage (AD), positive expiratory pressure (PEP), oscillating PEP, and high-frequency chest wall oscillation (HFCWO). With the exception of PEP, all techniques rely on the production of chest vibrations to speed up the removal of residual secretions.

The Cochrane collaboration confirms that regular use of ACTs by cystic fibrosis patients increases mucus transport in the airways, which significantly improves quality of life. [7] A one-year study was conducted in the UK to determine which airway clearance technique is the best. The study was conducted on a group of 75 patients with cystic fibrosis, 22 of whom did not complete the study. The participants were divided into 5 groups. The following techniques were compared: active cycle breathing techniques (ACBT), positive expiratory pressure (PEP), autogenous drainage (AD), and the oscillating PEP Flutter and RC Cornet devices. The study results did not show any statistically significant differences in the patients' health parameters. Each of them brings comparable benefits, so the patient should choose a technique that is convenient for him. [8] Studies have shown that patients are much more likely to choose to perform automatic drainage compared to postural

drainage and percussion (PD&P). [9] These procedures do not require additional people, the patient can perform them independently with the use of appropriate devices.
Additionally, in patients with cystic fibrosis, exercise training can be classified as ACT (airway clearance technique). For patients, it is often a less mentally burdensome technique of coping with respiratory ailments.Currently, exercise has been introduced in New Zealand and Australia as a therapy in the treatment of CF, which has a promising effect on the development of this trend in the treatment of the disease.[10]

Nutrition

For patients with cystic fibrosis, improved nutrition was the most important factor that extended the median life of patients by an additional 10 years. [11] In this group of patients, a more high-calorie diet is used in relation to the needs of healthy people. At the same time, techniques that improve the absorption and supplementation of vitamins and minerals should be used. [12]
In North America, a 10-year study was conducted to determine whether and how diet affects the development of patients with cystic fibrosis. For this purpose, in two clinics: Toronto and Boston, selected two groups of patients similar in terms of size, age, gender, ethnicity, lung function, etc. The diet of the first group was high-calorie with up to 30 capsules of pancreatic enzymes the other group's diet was low-energy with fewer capsules. [13] The results of the study clearly confirmed that the patients from the Toronto clinic were taller and gained more weight than the patients from the Boston clinic. Moreover, the median life expectancy of the patients was 10 years longer in the Toronto patients. Despite the fact that recently more and more attention has been paid to the importance of maintaining an appropriate diet in patients with cystic fibrosis, still not all patients achieve proper growth and nutritional status.
The Cystic Fibrosis Foundation (CFF) Registry, after conducting a study on 3,000 patients, states that at the age of 4, a higher percentile of weight in relation to age conditioned a higher survival rate of patients, their better condition of the respiratory system, and a higher BMI at the age of 18. [14]
Despite the fact that recently more and more attention has been paid to the importance of maintaining an appropriate diet in patients with cystic fibrosis, still not all patients achieve proper growth and nutritional status.

Oxygen therapy

Respiratory symptoms are among the most common and severe symptoms of cystic fibrosis. Undoubtedly, oxygen therapy significantly reduces ailments such as shortness of breath. So it became a concern whether chronic use of oxygen therapy would produce long-term effects or a cure. Zinman 1989 published a study of 28 patients evaluating the effect of night-time oxygen therapy on survival. (14 patients supplemented with oxygen during the night, the remaining 14 were in the placebo group). In both groups, 4 people died. The introduction of chronic oxygen therapy has not had a noticeable effect on the life expectancy of patients with cystic fibrosis. [15]
In a review, The Cochrane Collaboration describes that oxygen supplementation with polysomnography significantly shortens the period of falling asleep in patients. In turn, oxygen therapy during physical activity extended their possibilities. [16]

There is insufficient evidence to determine the effect of long-term oxygen therapy in the treatment of cystic fibrosis. Additional research is needed. The reduction of negative symptoms after the use of oxygen therapy in this group of patients is a promising harbinger of further research results.

Conclusion

Recently, the knowledge about the ways of treating cystic fibrosis has expanded, which significantly increased the median survival of patients. However, there are still about 80,000 people in the world who suffer from cystic fibrosis and will die prematurely due to its complications [17] Information on additional treatment methods needs to be deepened. There are few studies to compare these methods. There is no doubt that the treatment of cystic fibrosis should be multifaceted.

Depending on the severity of the disease, its treatment may vary, but the most important are: conservative treatment, symptomatic treatment and supportive treatment. Conservative treatment in cystic fibrosis consists in preventing infections by, for example, annual vaccination against influenza. Symptomatic treatment consists mainly in the treatment of ailments from the respiratory system, but also dysfunctions of the pancreas, liver, bile ducts or reproductive system. Supportive treatment includes techniques of purification of the respiratory system, oxygen therapy, as well as nutrition adapted to the needs of the patient. Simultaneous multi-system operation allows not only to increase the comfort of life of patients, but also to extend their life expectancy.

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