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# Urządzenia drenażowe wykorzystywane w rehabilitacji pacjentów chorych na mukowiscydozę

**Streszczenie**

Mukowiscydoza jest przewlekłą i nieuleczalną chorobą, wywołaną przez mutację genu kodującego kanał jonowy CFTR, który zapewnia prawidłowe nawodnienie śluzu gruczołów nabłonkowych (95% wody u osób zdrowych). W przypadku stwierdzenia mutacji, dochodzi do znacznego zagęszczenia śluzu w wielu narządach organizmu, w szczególności układu oddechowego i pokarmowego. Stosowanie aktywnych technik oczyszczania oskrzeli z wykorzystaniem urządzeń drenażowych stanowi jeden z najistotniejszych elementów kompleksowego postępowania fizjoterapeutycznego. Współcześnie zaleca się jak najwcześniejsze nauczanie praktycznego zastosowania urządzeń drenażowych podwyższających ciśnienie wydechowe lub zmieniających ciśnienie w świetle drzewa oskrzelowego tj: Maska PEP, Flutter czy Acapella, które oprócz wysokiej skuteczności terapii, zapewniają samodzielne funkcjonowanie pacjenta w życiu społecznym, bez uzależnienia od osób z zewnątrz.

**Słowa kluczowe:** mukowiscydoza, transport śluzowo-rzęskowy, oscylacja ściany klatki piersiowej.

# Application of drainage devices in rehabilitation of patients suffering from cystic fibrosis

**Abstract**

Cystic fibrosis is a chronic and incurable disease, caused by a mutation of a gene, which codes the ionic canal CFTR, which ensures correct hydration of the mucus of the epithelial glands (95% of water in healthy people). In the case a mutation is detected, a considerable concentration of mucus occurs in numerous organs of the organism, in particular the respiratory and digestive systems. Applying active techniques of bronchi cleaning with the use of drainage devices constitutes one of the most important elements of comprehensive physiotherapeutic procedure. Nowadays, it is recommended to teach, as early as possible, to practically use drainage devices increasing the expiratory pressure or changing the pressure in the lumen of the bronchial tree, i.e.: PEP mask, Flutter or Acapella. Apart from the high efficiency of the therapy, they provide independent functioning of the patient in social life, without the dependence on people from the outside.

**Keywords:** cystic fibrosis, mucociliary transport, chest wall oscillation.

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## INTRODUCTION

Within the last years, thanks to the dynamic progress in treatment and diagnostics, cystic fibrosis is no longer considered a paediatric disease [1]. The success connected with longer life expectancy and better functioning of patients could not be possible without the regularly performed physiotherapeutic procedures, enabling the thick mucus sputum, retained in the bronchial tree of a patient suffering from cystic fibrosis, to be evacuated [2,3]. In the long-term treatment process, the use of drainage devices in respiratory therapy (due to the high efficiency and prophylactic nature of the disease aggravations) should be considered as an activity which is necessary and obligatory. Currently, in respiratory clearance, the drainage devices are used, allowing therapy, which engages the patient to the maximum and brings uniform therapeutic benefits, as traditional postural drainage [4]. The most often used devices, recommended by the International Physiotherapist Group, are, inter alia: PEP mask, Flutter, Acapella and drainage vest [5].

### Positive expiratory pressure therapy – PEP system

In 1970 in Denmark the therapy applying the phenomenon of positive expiratory pressure in the airways was used for the first time. Since then, it has been successfully used in patients born with cystic fibrosis all over the world.

The aim of producing the condition of increased expiratory pressure in the chest is dilation of the distal sections of the bronchial tree. Achieving patency of the smallest bronchioli facilitates sputum evacuation outside and prevents the unfavourable phenomenon of bronchioli falling in [6]. The device used in the PEP therapy is an especially built mouthpiece or anaesthesiological-type face mask. It has a valve which causes resistance to the expiration and prevents regression of the expired air wave, which enables to achieve increased pressure inside the chest [7,8]. In addition, the device is equipped with a resistor indicating the current resistance value during the active inspiration at an average lung volume. The dosimeter allows managing an appropriate pressure level, which should be equal to 10-20 cm $H_2O$ [9]. The PEP mouthpieces can be also used by the patients as an adapter for aerosol-therapy procedure [2].

The modification of the PEP therapy is the Hi-Pep method prepared in Austria (High-pressure PEP therapy). In its application, the expiratory training is performed on the entire excessive expiration, and the resistance value reaches 40 to 100 cm $H_2O$. Repetition of strenuous expirations with resistance, performed through the PEP mask, most often leads to coughing reflex, which is a physiological reaction supporting coughing out the retained sputum outside [10] (Table 1).

Period of the therapy with the use of the PEP mask varies and depends on the amount of the evacuated sputum. The suggested breathing training for the patient is a session applied 2-3 times per day for, approximately, 15 minutes [6]. What needs to be taken into consideration during the therapy is good adherence of the mask to the face and preventing getting the air to the cheeks, which considerably decreases the efficiency of the exercise [11].

The most often used devices, applying positive expiratory pressure, are, among others: PARI PEP, Thera PEP, Resistex [12].

### Oscillatory positive expiratory pressure therapy – OPEP system

A continuation and supplementation of the PEP therapy is a method coming from Switzerland, which is based on the same phenomena, however, additionally a vibrating factor is applied in it, which is moved to the respiratory track walls. This was called the method of Oscillatory Positive Expiratory Pressure Therapy. The results of long-term research conducted all over the world confirmed that the oscillations, which make the bronchial tree vibrate, not only contribute to easier separation of thick mucus and its defragmentation, but also decrease its viscosity. This results in the fact that moving the retained sputum and its transportation to the upper sections of respiratory tract becomes much easier [2].

Currently, in the United States, there are available three types of devices producing positive expiratory pressure. These are the following: Quake, Flutter and Acapella from which the most popular and available are the two former ones [10].

Flutter has been known since 1994. It is a small, mobile device in the form of a conical tube with a carved funnel in which a steel ball is located. The air flow during the expiration performed through the mouthpiece and the change of the pressure it causes, makes the metal ball move in an oscillating way, with the frequency similar to the physiological ciliary movements (8-25 Hz). Quick changes of the pressure, as the effect of the ball ballottement, result in vibration of the bronchial tree, thanks to which the plugging mucus deposits are separated more easily. Their moving to the upper sections of the respiratory tract is facilitated by a long, deep expiration through Flutter, with producing positive expiratory pressure inside the chest (value approx. 5-35 cm $H_2O$) [2].

**TABLE 1. Advantages and disadvantages of the PEP mask.**

| PEP mask | |
|---|---|
| Advantages | Disadvantages |
| 1) Small device, simple in use | 1) It is not recommended in patients with dyspnoea |
| 2) After the principles are learnt, it requires no assistance of other people | 2) Possibility of occurrence of bronchospasms in patients with bronchial hyperreactivity |
| 3) Relatively low therapy costs | |
| 4) It can be applied in all age groups (infants, children, adults) | |
| 5) PEP mouthpieces can be used for nebulization | |

**TABLE 2. Advantages and disadvantages of Flutter.**

| Flutter | |
|---|---|
| Advantages | Disadvantages |
| 1) Small device, simple in use | 1) In advanced stages of the disease, the device can be ineffective |
| 2) After the principles are learnt, it requires no assistance of other people | 2) Wrong inclination of the device by the exercising person may influence lower training efficiency |
| 3) Relatively low therapy costs | 3) Applied only in older children and adults who can cooperate actively |
| 4) The method is tolerated by most of the patients | 4) There is a necessity to remove the mouthpiece during inspiration |

The patient is advised to perform 3 series 15 expirations each during one day (which takes approximately 15-20 minutes). Efficient use of Flutter requires a long training and concentration of the patient, who, thanks to his or her individual feelings, should place it in the most optimum angle [2,7]. Its slight moving up enables the increase in the produced pressure and frequency of vibrations, whereas placing it in a lower position produces a reverse effect. During the training, it is necessary to prevent the air from getting to the cheeks, which must be constantly stabilized by the patient, losing, at the same time, as small as possible amounts of the transmitted vibrations [2].

Another device that produces oscillating positive expiratory pressure in the chest is an American invention, so-called Acapella. Its advantage over Flutter consists in the possibility to set 5 different modes, enabling automatic change of the frequency and the pressure, which can be adjusted to the current health status of the patient [13]. The expired air is resisted by a resistor and the vibrations (0-30 Hz) are produced by a special valve, which interchangeably closes and opens the terminal outlet of the device. In addition, it is possible to perform the inspiration without putting the device away from the mouth as it was in the case of Flutter [2]. The inclination of the device during the training is irrelevant and does not affect the therapy effects [14]. Training with Acapella should last for 10-20 minutes in each of the two sessions during one day.

**TABLE 3. Advantages and disadvantages of Acapella.**

| Acapella | |
|---|---|
| Advantages | Disadvantages |
| 1) There is a possibility to set the resistance value of the expired air | 1) Price |
| 2)The device, irrespective of the inclination in relation to the ground, is effective in the same manner | |
| 3) It can be used in various positions, also drainage ones | |
| 5) There is no necessity to remove the mouthpiece during inspiration | |
| 6) There is a possibility to join the device with a nebulizer | |
| 7) It is easy to keep the device clean | |

Therapy with chest oscillations of high frequency – Drainage vest

The newest device offered in the therapy of respiratory tract clearance in cystic fibrosis patients is a drainage vest (HFCWO – high frequency chest wall oscillation), known as the so-called "the Vest" system [15]. It follows from the data prepared by the Cystic Fibrosis Foundation in the United States in 2010 that even 59.4% of American patients use it as the main therapy [12,16].

The research assessing the satisfaction and approach of patients to the applied therapy has shown that 50% prefer application of the drainage vest, not flutter, mostly driven by its higher efficiency [17]. A different opinion is popular in Canada, where other forms of therapy than the vest are much more often applied. The experts' reports as of 2009 confirm that almost 2/3 of Canadian patients at the age of 6 and more choose PEP as the main method supporting respiratory rehabilitation. Drainage vest is used much more rarely than in the United States, mostly due to the high cost of the device (approx. 1,600.00 American dollars) [12]. In European countries it is a new form of therapy that gains more and more recognition among the patients. The operation of this appliance consists in producing transitory air pulsations by a compressor, to which an inflated pneumatic vest is attached. The patient puts it on, at the same time embracing the chest from each side. The irregular, intermittent air flow results in its immediate, variable pressing of high frequency (HFCWC High frequency chest wall compression) or strong oscillations of the air which is inside the airways (HFCWO High frequency chest wall oscillation) [18,19]. For comparison with other methods, the values of the produced pressure reach 50 cm $H_2O$ at the frequency of approximately 25 Hz [7].

**TABLE 4. Advantages and disadvantages of a drainage vest.**

| Drainage vest | |
|---|---|
| Advantages | Disadvantages |
| 1) It causes simultaneous vibrations and oscillations of all sections of the respiratory tracts | 1) High price |
| 2) It can be applied in all age groups (infants, children, adults) | 2) The device takes more space than the other ones |
| 3) There is a large assortment of various types of vests which meet particular clinical requirements | 3) It can require assistance of another person |
| 4) They can be used in various positions, also drainage ones | 4) There are the following contraindications: haemoptysis, advanced stages of the disease, head, neck and chest wounds |
| | 5) Patients in the advanced stage of the disease may feel a considerable pain in the chest, therefore in these cases it is not recommended |
| | 6) It is not well-tolerated by some patients |

The producers recommend that, during the therapy, an adult person should keep the drainage vest for a maximum of 30 minutes in a standing or sitting position. The procedure starts from setting a lower pressure and lower frequency and, with time, increasing them to the recommended values [9]. Unfortunately, the main obstacle and impediment in popularization of this modern method are its costs, which are several times higher than those of other appliances used in respiratory rehabilitation.

It must be remembered in the selection of the forms of therapy, that using the vest is not always well tolerated by the youngest patients. During the research in Royal Brompton Hospital in Great Britain even six of ten patients considered the drainage vest to be uncomfortable and their feelings during the therapy were not positive [20]. In the situation of feeling bad, the appliance has a special button which, when pressed, enables to break the therapy.

## CONCLUSIONS

The explicit development of modern drainage devices in the last years is the result of searching for the best method supporting airways clearance, which would be both effective and comfortable for patients. Currently, in European countries, devices such as Acapella, Flutter, PEP mask

or drainage vest are considered an inseparable and essential assistance both in slowing down the disease progression and in obtaining life independence by children and youth suffering from cystic fibrosis [21]. Administration of pharmacological drugs (as the main therapy) preventing numerous infections of the respiratory tract is largely a result of insufficiently popularized new rehabilitation methods and too small number of specialists who are well – trained in this area.

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