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Review Article

Current Airway Clearance Techniques in Cystic Fibrosis

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Abstract

Pulmonary rehabilitation is the non-pharmacological treatment method aimed at alleviating symptoms, enhancing exercise capacity and improving health-related quality of life. It specifically acts in cystic fibrosis by improving sputum clearance, stimulating patients to be more physically active and self-confident. Physiotherapists play a central role in this process by delivering to the patients and actively teaching them physical exercises, a suitable method of airway clearance technique, and by encouraging self-adherence, a key action towards short and long-term results. There are fragile and dependent children, complex debilitated adults by multiple comorbidities, all of them needing complete assessment, correct follow-up and warm support. They will be mentioned the current used airway clearance techniques, together with their indications, limits and sometimes side effects.

Keywords: Cystic Fibrosis; pulmonary rehabilitation; Airway Clearance Technique; Positive Expiratory Pressure; High-frequency Chest wall Oscillation

Abbreviations

ACT: Airway Clearance Technique;

CF: cystic fibrosis;

CFTR: cystic fibrosis transmembrane conductance regulator;

PR: pulmonary rehabilitation;

PEP: positive expiratory pressure;

HFCWO: high-frequency chest wall oscillation

Introduction

Cystic fibrosis is one of the most life-limiting autosomal recessive diseases among people of European heritage. It affects one out of every 2000-3000 newborns in European Union, being more frequent in Northern European people, excepting the Finish one. It is present in 1 of 2500 live births in the UK [1] and it is less spread among African and Asian populations. There is therefore an understandable interest in early diagnosis, correct classification of cystic fibrosis transmembrane conductance regulator (CFTR), identification of CFTR related diseases, pharmaceutical treatment and referral to pulmonary rehabilitation (PR) programmes. The main objectives are to describe the current methods of airway clearance techniques (ACT) used in people with CF and the baseline characteristics for users of different ACT.

There are > 1800 CFTR gene mutations that can be classified based on the mechanism to generate disease. The mutation of F508del, deletion consisting in three nucleotides with the loss of the amino acid phenylalanine at residue 508, accounts for approximately 70% of all defective CFTR alleles, has a decreasing prevalence from north-west to south-east in Europe. This is the reason why in the UK there is a national screening programme where neonatal heel-prick trypsinogen measurement is offered routinely. The positive samples are tested for common CFTR mutations, sometimes followed by a second immunoreactive trypsinogen screen and by sweat testing [1].

Current Airway Clearance Techniques Used In Cystic Fibrosis

CF is one of the five obstructive pulmonary diseases where PR is highly recommended, next to chronic obstructive pulmonary disease, persistent bronchial asthma, diffuse bronchiectasis and bronchiolitis obliterans [2]. Patients are referred to PR for persistent respiratory symptoms (dyspnea, fatigue, cough and persistent sputum) and/or functional status limitation despite an optimal pharmacological treatment. The wisdom in sending people to PR, draw from multiple clinical studies, is to make this recommendation at a milder stage of disease, when the accent will drop on preventive strategies and efforts to maintain physical activity [2]. PR reduces symptoms, enhances sputum clearance, increases exercise capacity and improves quality of life in CF patients with high variable needs.

From the very complex PR team, a central role play physiotherapists, having a key position in the assessment of functional ability with exercise tests, evaluating treatment and encouraging aerobic training [1]. They effectively teach ACT in order to be performed by the patients themselves twice daily on a long-term basis. Despite the proven reality that attending PR courses accompanied by someone in the family will become a stimulus to train at home, it is more difficult with the children because they will be convinced to practice only the exercises that will be accepted by the parents/tutors, and that might not be the most recommended strategy [3].

After the changes made to the ACT categories in 2013 by the CF Trust, people in Britain are registered to use one the following primary ACT: postural drainage, forced expiratory techniques (including huffs, active cycle of breathing techniques and autogenic drainage), positive expiratory pressure (PEP), oscillating PEP (e.g. flutter valve and Acapella), high-frequency chest wall oscillation (e.g. vest), other (usually non-invasive ventilation), exercise and non-specified. This information was withdrawn from the 2011 UK CF registry [3]. Active cycle of breathing techniques means: tidal volume breathing, then deep inspiration and passive expiration, followed by forced expiration to mobilize secretion prior to coughing/huffing [1].

Hoo et al have found that the most common used ACT in the UK in 2011 were forced expiratory techniques and oscillating PEP, whereas postural drainage and HFCWO were obviously less preferred by the patients. Also, the male:female ratio of individuals who used exercise as their primary ACT was 2:1, compared with 1:1 for other techniques [3]. Up to this point, there is a small interest of patients in physical exercises (nothing new here), as they prefer to have some sort of technique to have things done for them. Also, females are ashamed to spit; physical exercises increase mucociliary clearance peripherally [4] and stimulate sputum to move out, apparently facing a gender limitation.

Still, things are not so easy! Traditional ACTs are relatively cumbersome and time-consuming to perform. Postural drainage concerns through the head-down tilting that may exacerbate gastro-oesophageal reflux and dyspnea. It is also mentioned that while using HFCWO, forced expiratory flow 25% to 75% (FEF25-75) has decreased comparing to other techniques, suggesting a potential mismatch between objective and perceived efficacy. So here we come to the patient preference and self-reported adherence, essential to make the selected ACT to work optimally and create self-reported satisfaction. Also, McIlwaine et al have reported several studies where various ACTs have same efficacy on airway clearance in maintaining health in CF patients, without any superiority of one of them. But one study suggested that HFCWO was not as effective as PEP in maintaining health in CF patients [4].

Using the UK registry, it was shown that 15% of British people use exercise as their primary ACT, and sometimes unique, despite recommendations to play only an adjunctive role in the ACT arsenal [3]. It is advisable to psychological assess this liability to physically exercise, a non-stigmatizing activity performed by all healthy-people, with numerous other benefits on physical and general well-being. This is in line with the recommendations made by Osadnik et al that clearly emphasize how beneficial is PR for many respiratory patients groups, including those with CF [5]. Skilled healthcare professional

should recognize that people with respiratory conditions, especially those with chronic diseases, are less physical active than healthy individuals and physiotherapists should induce behavioral changes to facilitate adoption and maintenance of an active lifestyle. Pulmonary rehabilitation should target both psychological well-being and self-management of the patient's respiratory condition.

Moreover, children and adolescent with CF present postural changes in comparison to healthy peers. Physical exercises consisting in aerobic exercise and stretching help to improve posture and prevent progression of some postural disorders. In a recent study published by Schindel and coworkers, an educational guideline for exercise practice have reduced cervical lordosis, thoracic kyphosis, lumbar lordosis, lateral chest distance, abdominal protrusion, the baropodometric mean pressure and contact area in individuals aged 7 to 20 years [6]. Besides, CF patients that undertake regular aerobic exercises maintain higher indices of respiratory muscle strength and lower pressure-time index of the respiratory muscles, as proved by Dassios et al [7].

It is quite clear that among oscillating devices, British people pay attention rather to the oscillating PEP, being less attracted by the HFCWO vest. After a pertinent comparison of several studies on chest physiotherapy in CF, Morrison and Agnew [8] concluded that there is no clear evidence that oscillation technique is more effective than any other form of physiotherapy; also, no oscillating device is superior to the other one. It is a matter of good device choice, where adherence to a certain technique for a long-term intervention on airway clearance will also improve exercise tolerance and respiratory function. It might also be a matter of money: a PEP mask costs approximately 50 pounds, and a HFCWO device is 7000 pounds [9].

Let's also consider that when breathing out against resistance in order to achieve positive expiratory pressure, the underlying physiology of PEP allows multiple benefits to be reached like increased lung volumes, decreased hyperinflation or improved airway clearance [10]. Clearance is improved by building up gas behind mucus via collateral ventilation and by temporary increasing functional residual capacity [11].

McIlwaine et al spoke as well on adherence to a certain oscillating method, stretching out that perceived effectiveness of a treatment may lead to improved adherence and treatment satisfaction. They found that there are no significant differences between PEP and HFCWO. Patients have appreciated more the PEP technique for flexibility and a shorter treatment time. But there were more adverse events related to the lower airways in the HFCWO group as increased cough, chest infections, hemoptysis, decreased lung function and chest pain [9]. Data support the use of PEP therapy as the primary ACT in CF patients aged > 6 years; the study does not support the use of HFCWO as a primary ACT in CF patients.

The review published by the same authors in 2015 [11] has considered as the primary outcome being the forced expiratory volume in one second, and a second primary outcome the number of respiratory exacerbations. There was declared a lower exacerbation rate in participants using PEP comparing to other technique when using a PEP mask for at least one year.

Conclusions

The patient's preference exerts a great influence on the choice of the ACT. Instead of the old postural drainage, the "gold standard" of past times, today CF patients prefer ACTs that can be self-administrated in any time and sometimes in any position, like: forced expiratory techniques, oscillating PEP and PEP. Nowadays PEP has become the "gold standard" in the physiotherapy of cystic fibrosis. People with mild lung disease are prone to use exercise alone; people with more severe disease prefer HFCWO and nebulized mucolytic/osmotic; that means that some techniques as HFCWO and non-invasive ventilation should be reserved to sicker patients [3]. Making a good technique choice is a key action to meet patient's preference and to ensure short and long-term adherence and also the results of the intervention.

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