



Title: Clinical Implications High Frequency Chest Wall Oscillation (HFCWO)

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Abstract

Purpose: patients with neuromuscular diseases presents an high incidence of respiratory infections favoured by stagnation of deep bronchial secretions and deficit of cough. The aim of the study is to evaluate the correct treatment of this condition and the role of High Frequency Chest Wall Oscillation (HFCWO) in helping the removal of bronchial secretions and reduce the incidence of infections in patients with neuromuscular disease.

Methods: analysis of the current bibliography related to respiratory infections and neuromuscular disease. PCEF (Peak Cough Expiratory Flow) is used as a standardized indicator of efficiency of cough.

Results: the High Frequency Chest Wall Oscillation (HFCWO) is useful, in cases of increased production of mucus and impairment of muco-ciliary clearance, to remove the tracheobronchial secretions and reduce the incidence of infections.

Conclusions: the correct approach to patients with neuromuscular disease and frequent respiratory infections is focused on treatment of cough ineffective and management of bronchial secretions. High Frequency Chest Wall Oscillation (HFCWO) (VEST) has a central

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role in treatment of cough ineffective and management of bronchial secretions reducing respiratory infections.

1. INTRODUCTION

Respiratory infections are the most common cause of hospitalization and the most severe complication of patients with neuromuscular diseases. The most important reasons for this condition are the stagnation of deep bronchial secretions and the usual presence of a deficit of cough. The PCEF (Peak Cough Expiratory Flow), the most important standardized indicator of efficiency of cough, as a matter of fact, is always reduced. The consequence of this condition is a decreased of the tracheobronchial secretions clearance with a predisposition to infection. Moreover, the reduced physical activity, the high frequency breathing and the low tidal volume gradually lead to an increase of stiffness of the chest wall until sternum chondral joints ankylosis. The reduction of PCEF in neuromuscular diseases is caused by several factors. The most important are the weakness of respiratory muscles and the reduced compliance of chest wall and lung. The reduced expansion leads to the formation of pulmonary atelectasis of the lung parenchyma and infectious episodes. Among the most commonly used spirometric parameters, vital capacity (VC) is reduced, even if, the value must be considered significant when it is equal to or less than 50% of normal. The maximal inspiratory pressure (MIP), maximal expiratory pressure (MEP) and the nasal inspiratory pressure (SNIP) are also reduced. The value of PCEF, directly represents the effectiveness of cough. The value of PCEF considered normal in adults is greater than 360 L / min. Some authors consider worth enough for an effective cough a value of 160 L / min. Other authors sustain that to overcome a bronchopulmonary infection is needed a PCEF value bigger than 270 L / min (Table 1).

2. PHYSIOLOGY OF COUGH

The cough is an automatic mechanism of defence. It however, can be reproduced and partially controlled voluntarily. It is especially useful in cases of increased production of mucus and / or impairment of muco-ciliary clearance to remove the tracheobronchial secretions. This mechanism is called "cough clearance". It use the power of a forced expiratory to clean airways from irritants and secretions. The reflex is triggered by stimulation of receptors placed in the airway: larynx, trachea and bronchial tree. There are also extrapulmonary receptors located in the pleura, esophagus and external ear. The afferent pathways are constituted mainly by the vagus nerve (larynx, trachea, carina, pleura, bronchi), the glossopharyngeal (pharynx) and the intercostal



nerves. The centre of the cough is located close to the breath centre, in the medulla oblongata. The efferent pathways are represented mainly by the inferior laryngeal nerve (glottis), the phrenic nerve (diaphragm) and spinal nerves (intercostals and abdominal muscles). The cough is the final phase of a reflex that can be divided in four stages: irritation, inspiration, compression and ejection (Table 2):

1. *Irritation*: stimulation of cough receptors placed in laryngo-tracheal and in bronchial tree.
2. *Inspiration*: it must be rapid and deep to facilitate the return of elastic lung parenchyma, with the aim of producing the maximum expiratory flow possible. High lung volume optimizes the voltage-length of the expiratory muscles, allowing to generate high pressures and high expiratory flow. During this phase, the glottis is open thanks to the abductor muscles of the cartilages aritenoidee. This allows the divergence of the vocal folds allowing air to enter quickly in the lung.
3. *Compression*: strong expiratory with closed glottis. Its duration is about two seconds. Intrathoracic and abdominal pressure considerably increase due to the contraction of expiratory muscles of the chest, abdomen and pelvic floor.
4. *Expulsion*: is the classic cough. Following the sudden opening of the glottis and the concomitant elevation of the soft palate. This phase is characterized by an explosive release of air at high flow and a high-frequency vibration of the bronchial wall. As a result of the sharp fall in intrathoracic pressure, the diaphragm is passively propelled upward, the air is violently expelled, and the intrapulmonary bronchial material dragged out. The diaphragm during the third and fourth phase is released and the intra-abdominal pressure is transmitted to the thoracic compartment. In healthy individuals the air intrapulmonary expiratory flow reaches more than 6 l / sec. The increased speed of airflow, as determined by the reduction in size of the airways, allowing contact between air and mucus layer, known as "two-phase flow" (gas and liquid). The posting of secretions is possible due to the transfer of kinetic energy from molecules of air to those of mucus. The gas-liquid interaction between the air at high speed and large molecules flow of mucus is the mechanism of removal and transport of secretions. The action of coughing is expressed at the level of airway proximal to the 6[^] -7[^] bronchial generations. It represents one of the more effective mechanisms of tracheo-bronchial toilet.

3. NEUROMUSCULAR DISEASE: RESPIRATORY FISIOPATOLOGY



Neuromuscular diseases (Table 3) are an heterogeneous group of diseases, but independently from the different etiopatogenesis and clinical features have a similar natural history with similar stages of evolution. The following conditions are observed in all neuromuscular diseases: a reduction of the strength of respiratory muscles, a restrictive respiratory syndrome and a deficit of cough with stagnation of secretions in the airways and consequent atelectasis.

3.1 - REDUCTION OF THE STRENGTH OF RESPIRATORY MUSCLES

These patients have a decrease in inspiratory and expiratory muscle strength expressed by lower peak inspiratory pressure (MIP) and maximal expiratory pressure (MEP). In spinal cord injury patient with medium-low level of lesion (C4 - C7) the reduction of MIP in the acute phase is $64 \pm 12\%$ but increases about 40% (46 ± 6 to 77 ± 4) within 18 weeks .

3.2 - RESTRICTIVE RESPIRATORY SYNDROME

The reduction in lung volumes measured by spirometry, is mainly due to lack of ventilatory pump, but also to the alterations of the mechanical properties of the chest and to the formation of atelectasis. The vital capacity is always reduced. With values of vital capacity around 40-50% of normal ventilatory pump deficiency becomes constant and particularly during infection is necessary medical treatment. The inadequacy of the ventilatory pump can be maintained or worsened by:

- Overproduction and stagnation of secretions favoured by the deficit of expectoration.
- Increased load and resistance of airways. The reduced muscle strength and alteration of the elastic properties of the respiratory system prevent the achievement of adequate lung volumes after an in-depth. This limits the amount of air available to the expulsive phase of cough and indirectly causes a decrease in expiratory flow and, in particular, the PCEF.
- Further reduction of lung compliance for the formation of atelectasis and infections.

In case of quadriplegia all lung volumes are reduced, except the residual volume increases by reduction of reserve capacity expiratory. The FRC is reduced: this parameter is an expression of balance between the opposing elastic forces of lungs and chest from which it derives a new equilibrium at lower lung volumes. FEV₁ (maximum expiratory volume per second) is reduced with values similar to the reduction of vital capacity. The stagnation of secretions generates an increased work of breathing worsening respiratory mechanics and increasing muscle fatigue leading to a serious deficit in ventilatory pump. Another major complication is represented by atelectasis (mainly in the dorsal lung areas) also in small bronchi of the deep lung. The weakness



of expiratory muscles reduces the magnitude of expiratory flow with consequent alteration of the expulsive phase of cough and further stagnation of secretions and possible pulmonary atelectasis

4. PRINCIPLES OF TREATMENT

The main objectives are the treatment of cough ineffective and the management of bronchial secretions.

4.1 Treatment of cough ineffective.

The effectiveness of cough may be impaired by alterations of a single specific phase of cough. After a specific evaluation is correct to adopt the most appropriate technique. The rehabilitative intervention is, therefore, depending on the stage of cough in which there is the deficit. The deficit can affect the phase of inspiration, compression and / or expulsion. Phase of inspiration: when the CV is less than 1500 ml. In this case the rehabilitative intervention will be aimed at increasing the volume inspired or pre-cough (Air or Mechanical Stacking In Ex-Sufflator). Compression phase: when the PCEF is less than the PEF, the MEP is greater than 40 and the PCEF is less than 3 liters per second. In this case, the appropriate rehabilitative care will be oriented to the manual cough (Air Stacking) or the use of In Ex-Mechanical Sufflator. Ejection phase, when the MEP is less than 40 cm H₂O and when the PCEF is less than 3 liters per second. In this case the treatment will be directed to the manual cough assistance (Air Stacking) or the use of In Ex-Mechanical Sufflator. In case of global impairment, both inhalation and exhalation technique may be used, such as re-expansion procedures, assistance coughing procedures or using the Mechanical In Ex-Sufflator. Air Stacking is correctly repeated for 3 cycles daily through facial mask or mouthpiece. Air Stacking, Mechanical In Ex-sufflator and nocturnal Non-Invasive Mechanical Ventilation induced an higher distensibility and elasticity of the chest wall and an increase of the MIC (maximum capacity insufflatoria) preventing the atelectasis (3).

4.2 Management of bronchial secretions.

Current recommendations for the management of tracheobronchial secretions in patients with respiratory muscle weakness involving the control of posture, physical therapy and use of modern technology such as the VEST. The control of posture through the variation of patient position in bed prevents the stagnation of secretions in order to facilitate their removal and improve gas exchange in all districts affecting the pulmonary regional distribution of pulmonary ventilation. In addition to the control of posture are important the techniques of physiotherapy and clapping. However, these procedures induced fatigue in all patients and there may be important episodes of



oxyhemoglobin desaturation. For this reason is important to consider alternative techniques that provide tools to reduce the muscular work for the mobilization of the patient's secretions. One of these new techniques considered particularly effective is the VEST.

5. Using of VEST in neuromuscular diseases.

The VEST with the vibrations transmitted to the chest can play a key role in helping the removal of bronchial secretions and reduce the incidence of atelectasis in patients with neuromuscular disease. The high frequency oscillations transmitted from the chest wall to bronchial secretions induced a more easily detachment from the wall and also a change of their qualitative characteristics. Secretions become more fluid and more easily removable. The parameters are set by in a not-standards way to differentiate the therapy in different patient to obtain a maximum tolerated pressure for every patient. However in our experience a successful plan follow a scheme with a vibration of 7-8 Hertz and a pressure of 6-8 cm H₂O for a period of about 10 minutes. The frequency and the pressure are increased gradually until reaching the maximum tolerated by the patient. The sessions last about 10 minutes and are repeated 3 times a day. In some patients we used treatments with different frequency and pressure during the day to put in resonance different bronchial and pulmonary structures. Patient who still has an effective cough can eliminate spontaneously and easier also the most profound secretions. Patient who are tracheotomised can be aspirated easily. Patient not tracheotomised with ineffective cough can use Mechanical In Ex-Sufflator after the treatment session with VEST or in a pause of the VEST treatment if the quantity of secretions mobilized becomes important. VEST and Mechanical In Ex-Sufflator have the advantage of not requiring any cooperation from the patient and can be used both in the acute phase - even during mechanical ventilation – and in the chronic phase. Their use can also be done to the patient's home being particularly simple to use and really easy the training of a care giver. In our experience the use of VEST together with MIE has become a classic instrument to prevent respiratory infection and has enabled a reduction of hospitalization in neuromuscular patients.

Table 1: Reference values of PCEF

PCEF > 360 L / m ' Normal in adults	<i>Bach JR et al.</i> Arch Phys Med Rehabil (1993)
PCEF < 160 L / m ' Clearance bronchial secretions insufficient (adult)	<i>Bach JR et al.</i> Chest (1996)
PCEF > 160 L / m '	<i>Mier-A Jędrzejowicz</i> Am Rev Respir Dis (1988)



Clearance for bronchial secretions insufficient during respiratory infection	
PCEF > 270 L / m '	<i>Bach JR</i> Chest (1997)
Value to indicate an effective cough	

Table 2: Phases of Cough

<i>Irritation</i>	Starting stimulus
<i>Inspiration</i>	Precough lung volume is reached through a depth inspiration
<i>Compression</i>	The contraction of expiratory muscles to generate high pressure with closed glottis
<i>Expulsion</i>	The opening of the glottis allows air flow at high speed to clean out these secretions from airways

Table 3: Neuro muscular

<i>I. Myopathies</i>	<i>II. SPINAL CORD DISORDERS</i>
Muscular dystrophies	Spinal muscular atrophies
Dystrophinopathies Muscular (Duchenne, Becker)	Motor neuron diseases
Limb-girdle	Poliomielitis
Emery-Dreifuss	Spinal cord injuria
Facioscapulohumeral	Transverse myelitis
Congenital	<i>III. Neuropathies</i>
Childhood autosomal recessive	Hereditary neuropathies
Myotonic dystrophy	Any condition with diaphragm paralysis
Congenital and metabolic myopathies	Guillain-Barré syndrome
Inflammatory myopathies	Multiple sclerosis
Diseases of the myoneural junction	Friedreich ataxia
Mixed connective tissue disease	<i>IV. OTHERS DISEASES</i>
Myopathies associated with systemic conditions	Familial dysautonomia, Down syndrome

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