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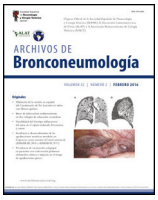

Editorial

Respiratory physiotherapy in Lady Windermere syndrome: The missing link?☆

La fisioterapia respiratoria en el síndrome de *Lady Windermere*: ¿el eslabón perdido?

Lady Windermere syndrome was first described in 1992 by Reich and Johnson.[1] It was named after the female character in Oscar Wilde's play – Lady Windermere – who hid behind her fan to voluntarily inhibit coughing, since it was not socially acceptable for "ladies" to clear secretions in public. This syndrome was described in non-smoking women, with a mean age of 60–70 years, no underlying lung disease, growth of *Mycobacterium avium* complex in respiratory samples, and development of bronchiectasis mainly involving the middle lobe and/or the lingula. Other phenotypic characteristics associated with the disease have been described, such as pectus excavatum, scoliosis, mitral valve prolapse, and a slender body habitus.[2] One of the etiological factors involved in its pathophysiology is the voluntary suppression of cough, which, coupled with anatomical peculiarities of both the middle lobe and the lingula and long narrow bronchi, would favor the accumulation of secretions in these lung regions. In addition, a slender body habitus, with a narrow anteroposterior diameter of the chest, increases this regional susceptibility, and together with pectus excavatum can limit the drainage of secretions. The existence of a genetic predisposition for the presence of allelic variants of the gene regulating the transmembrane conductibility of cystic fibrosis and its phenotypic similarity with inherited connective tissue disorders can also contribute to its pathogenesis.[3,4] Other factors such as alterations in systemic leptin or adiponectin levels and menopause may also play a role in pathogenesis.[5]

The presence of *Mycobacterium avium* complex usually precedes the development of bronchiectasis, so it is important to assess whether it meets criteria for treatment,[6] which is long (minimum 1 year) and complex due to the potential side effects of the drugs used. Initial combined treatment with clarithromycin/azithromycin, rifampicin, and ethambutol is the right approach in most patients. Given the high estimated health resources needed[7] and effects of long-term antibiotic treatment, the development of non-pharmacological therapies that support conventional treatments but do not cause adverse effects is being encouraged.

The latest national and international pulmonology guidelines[8,9] recommend drainage of secretions in patients with bronchiectasis from the early stages, so it is clear that respiratory physiotherapy (RP) would benefit these patients and hospitals need to make adequate provision for efficient RP programs with specialized physiotherapists. Indeed, a recent survey conducted in Belgium reported that up to 84% of patients with bronchiectasis were seen by a respiratory physiotherapist for bronchial hygiene.[10] Therefore, drug treatment aside, in patients with Lady Windermere syndrome, as in patients with bronchiectasis, the drainage of mucopurulent secretions that can favor infection or colonization by pathogenic organisms may be crucial to breaking the circle described by Cole et al. in 1986.[11] In this entity, as in any other non-tuberculous mycobacterial (NTM) infection, it is unclear whether it is the chronic NTM infection that causes bronchiectasis, or whether the previous presence of bronchiectasis facilitates colonization by NTM. Whatever the case, the inability to eradicate NTM from the bronchial tree causes a chronic granulomatous inflammatory response with increased inflammatory cytokines, chemokines, and neutrophil recruitment. These release proteolytic enzymes leading to destruction and damage of the bronchial wall, with the formation of new bronchiectasis. Moreover, this inflammatory cascade causes greater changes in mucociliary clearance, favoring the formation of mucous plugs and perpetuating chronic airway inflammation.[12] Thus, in this population with a high inflammatory burden and voluntary suppression of the cough mechanism, priority must be given to RF techniques that facilitate the drainage of secretions, although other techniques also exist.

In order to ensure airway patency, physiotherapists apply two types of techniques: manual and instrumental. The manual techniques are: autogenous drainage, total slow expiration with open glottis in lateral decubitus (ELTGOL, from the French Expiration Lente Totale Glotte Ouverte en décubitus Latéral), active ventilatory cycle, and cough itself; while the instrumental techniques are performed using positive oscillating and non-oscillating expiratory pressure devices, most often external high-frequency oscillating-compression devices on the chest wall and percussive intrapulmonary ventilation.[13] Treatment should begin with manual techniques that allow good bronchial hygiene to be maintained by modulating the expiratory flow (laminar or turbulent) depending on the location of the secretions. Thus, low expiratory (laminar) flow techniques will be applied for distal secretions, and as these move towards more central airways (assessed by respiratory auscultation), high expiratory (turbulent) flow techniques will

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be applied. This sequence of maneuvers will make it easier for the patient to finally be able to cough.

Scientific evidence currently underlines the utility of ELTGOL among bronchial hygiene maneuvers. ELTGOL involves the patient being be placed in lateral decubitus, on the affected lung, thus favoring maximum deflation and gas-liquid interaction. The patient is then asked to breath normally at tidal volumes, and then breathe out the air with the glottis open, producing a laminar flow until the end of the expiratory reserve volume is reached, preferably with the manual assistance of the physiotherapist.[14] In 2018, Muñoz et al.[15] provided more evidence on this technique with the publication of a randomized study which concluded that applying ELTGOL twice a day for 1 year not only produced an increase in the amount of sputum mobilized, but also reduced the number of exacerbations, improving quality of life in patients with bronchiectasis.

Today, as in the case of bronchiectasis, RP combined with long antibiotic treatments to reduce the inflammatory burden in the bronchial tree play a fundamental role in improving clinical management of patients with Lady Windermere syndrome. More randomized studies are needed to identify the effects of RP and the best bronchial hygiene techniques in these patients.

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