Effects of Mechanical Insufflation-Exsufflation on the Breathing Pattern in Stable Subjects With Duchenne Muscular Dystrophy: A Step in a Wrong Direction

To the Editor:

A recent study investigated the short-term effects of a mechanical insufflation-exsufflation (MI-E) device on lung mechanics and breathing pattern in subjects with Duchenne muscular dystrophy (DMD).[1] Cesareo et al[1] stated that "the primary outcome of the study was to evaluate the acute effects of a single treatment of MI-E in subjects with DMD who have no active upper airway or lung infection, in terms of lung recruitment changes, compliance of the respiratory system, breathing pattern, and volitional cough"; indeed, the results of the study showed an improvement in the rapid shallow breathing index. There were no changes in cough peak flow or vital capacity measured after treatment. However, the authors missed the most important point: the main goal of MI-E devices is to increase cough peak flow generated by the therapy and thereby increase cough strength and assist in secretion clearance. We understand that there is a need for physiological studies, but we are unsure what this study adds to the current literature base. Therefore, the purpose of this letter is to avoid confusion and highlight the main focus and outcomes of MI-E. We will also explain the possible misunderstandings that occur when MI-E is used in a manner that is not in line with the goals for which the device was designed.

First, the authors measured unassisted cough peak flow before and after the subjects used MI-E. It is hardly surprising that there was no increase in cough peak flow or lung volume because other studies have shown this in conjunction with the technique.[2] Based on their protocol, Cesareo et al[1] should have been able to provide information about the MI-E assisted cough peak flow and lung volumes because they carried out 5 cycles of MI-E, and these results are available with the fifth generation of MI-E devices.[3]

Second, MI-E was originally designed to assist cough during respiratory infections,[4] and it has been demonstrated that patients with neuromuscular disorders do not use MI-E routinely at home when they are stable.[5] MI-E is considered by an international expert group to be an expensive, but ideal, technique to assist cough in patients with severe DMD.[6,7] Non-ambulatory patients affected by DMD are likely to develop respiratory infections with declining lung volume and cough capacity.[8] Lung volume recruitment techniques providing deep lung inflation have been suggested to recruit volume by preserving lung and chest wall compliance when used once or twice daily.[8] Studies have investigated inexpensive techniques, such as breath-stacking using volume-preset ventilation or a manual resuscitator bag,[9-11] as well as expensive techniques, such as MI-E,[12,13] which has a cost nearly 100 times greater than that of breath-stacking via a resuscitator bag. It is not clear why Cesareo et al[1] decided to investigate the more expensive option of MI-E when less expensive techniques are available and effective. Their data show that 13 of 20 subjects used noninvasive ventilation (NIV) at night.[1] We therefore assume that subjects could use NIV rather than MI-E for daytime lung recruitment with no additional cost. It is hard to justify to patients, families, the health care system, or private health insurers that a MI-E device should be provided to maintain an unassisted cough as evaluated in this study. Is that a reasonable goal? We know that MI-E devices are effective in clearing airway debris that cannot be cleared with non-instrumental techniques.[4-6] Using MI-E to recruit lung volume appears to be an expensive way to reach this goal. The authors found a short-term effect on dyspnea and breathing pattern after using MI-E. What is the clinical message of this? Do the authors advise using MI-E to improve dyspnea and the breathing pattern? Surely the evidence-based consensus is that NIV is the best treatment option to achieve this goal in patients with DMD.[8,11,14]

Third, the authors described using MI-E pressure settings of 34 ± 5 cm H_2O for both positive and negative pressures[1]; however, a normal cough expels 85–90% of total lung capacity, a volume often unattainable at insufflation pressures < 50 cm H_2O.[4] We understand that positive pressure may recruit volumes, but there is little physiological explanation to argue that negative pressures will help maintain lung compliance and volumes. Using positive pressures solely could perhaps produce similar or better results than those measured with both pressures, and, possibly, positive pressure alone could be perceived by patients as more comfortable in such a study design.[1] We see similar changes in breathing pattern after a hyperinsufflation with mouthpiece ventilation.[14]

Fourth, we acknowledge that experts in respiratory care can understand the aim of such a study that provides a dubious clinical message, which was not highlighted in the limitations of this study. Unfortunately, non-experts are likely to misunderstand the lack of clinical impact of this study. In the past we faced people asking for MI-E devices for lung recruitment. However, with a better understanding of the intended use of MI-E devices,[7] requests for appropriate and intended device usage is improving. Studies like this one provide misconceptions about the objective intended use of MI-E devices. Therefore, we must explain to patients with DMD and their families that inexpensive techniques can provide results similar to those obtained with expensive techniques. Education on how to use MI-E in assisting cough is challenging. Occasional goals such as lung recruitment with MI-E distract people from the right goal and may result in misusing MI-E devices.

That said, we agree that patients with DMD who are living in areas where MI-E is available can achieve multiple long-term goals of cough augmentation, chest wall stretching, and lung volume recruitment. Our objective here is to remember that there is no reason to use a MI-E device in stable patients with DMD who have effective unassisted cough peak flow to specifically target lung volume recruitment. We also highlight the necessity to conduct studies investigating the preventive use of a MI-E device with regard to the rate of infections, exacerbations, and hospitalizations, as well as the benefits of a MI-E device in improving assisted cough peak flow values.[2]

The authors stated that the study reported the first validation of optoelectronic plethysmography in slow vital capacity. It is our opinion that the authors should have concentrated on this as their main outcome so as not to detract from the important clinical message that MI-E is used to clear secretions rather than to change breathing frequency. Moreover, according to the available evidence and our clinical experience of > 20 y working with MI-E, there are easier and more efficient ways to optimize MI-E settings based on the impact of achieving competent cough peak flow to overcome secretion encumbrance in patients with DMD.

In conclusion, the clinical messages of this study are not clear and may create confusion among clinicians who treat patients with DMD. Many clinicians are struggling to use optimal settings when using MI-E within the goals for which this device was designed. We believe that MI-E studies must focus on assisting cough rather than improving breathing pattern and lung mechanics.

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Effects of Mechanical Insufflation-Exsufflation on the Breathing Pattern in Stable Subjects With Duchenne Muscular Dystrophy: "A Step Into New Knowledge"

We thank Drs Toussaint, Gonçalves, and Chatwin for their interest in our paper.[1] However, we do not think that our results may in any way create confusion or change the main focus and outcomes of mechanical insufflation-exsufflation (MI-E) treatment. On the contrary, as underlined by Joshua Benditt in his editorial accompanying our paper,[2] we believe that our data add new information by analyzing "the physiologic effects on the respiratory system of the application of MI-E as a routine application and not for secretion removal" in patients with respiratory muscle weakness due to Duchenne muscular dystrophy (DMD). In addition, our data confirm that MI-E is not effective for purposes other than to assist cough, at least in late-stage, non-ambulatory DMD subjects with inefficient cough without ongoing airways infections.

Toussaint et al named two main concerns: to consider MI-E rather than other techniques or devices to assist cough, and to consider MI-E for lung recruitment. We are fully aware that the techniques to assist cough range from simple manual assistance (eg, using a manual resuscitator bag) to more expensive cough-assist devices. We are also aware that, according to the geographic area or the national health care system, patients may not have access to all of the aforementioned devices and could use less expensive techniques. In our paper,[1] we reported data collected in a tertiary care center in Italy where all DMD patients with ineffective cough have free access to MI-E devices, which are supported by the Italian National Healthcare System.

The median unassisted cough peak flow of our patients was 163.0 L/min with an interquartile range of 85.2 L/min. These values are far below the threshold of 270 L/min, which distinguishes efficient cough from inefficient cough[3,4] and is considered by all of the international guidelines for the management of patients affected by DMD.[5] This is an important point missed by Toussaint et al, who wrote in their letter, we should "remember that there is no reason to use a MI-E device in stable DMD patients

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