Considerations About the Effect of Cough Assist on Laryngeal Function in Neurologic Disease

To the Editor:

We read with interest the work by Andersen et al[1] about transnasal laryngoscopy during mechanical insufflation-exsufflation in amyotrophic lateral sclerosis (ALS). We believe the following should be considered.

First, the larynx is a complex structure that cannot simply be thought of as a passive valve through which one may insufflate and exsufflate the lungs with similar responses in all, particularly not in patients with intrinsic laryngeal weaknesses and reduced motor control. Given the nature of ALS, with its combination of upper and lower motor neuron signs, glottic narrowing in patients with ALS could be the infranuclear paralytic type, the supranuclear nonparalytic type, or a combination of the two. Vocal cord abductor paresis and laryngospasm in ALS are rarely described in the neurologic literature and autopsy studies.[2]

During exsufflation in all patients with neuromuscular disorders, including ALS, there was constriction of the hypopharynx, an exaggeration of the findings seen in patients without neuromuscular disorders to a varying degree. Constriction seemed very pronounced in subjects with hypotonic bulbar weakness. Clearly, airway closure on both insufflation and exsufflation is a major limitation in applying mechanical insufflation-exsufflation and is likely to be distressing in those patients most severely affected. Andersen et al[1] showed that adduction of supraglottic structures limits insufflation in patients with bulbar neuromuscular disease, including ALS. They surmise that upper airway reflexes may be hyper-regulated to prevent aspiration of foreign material into the airways, which will stimulate glottis closure, with the collapse of more floppy supraglottic structures.[3]

Second, mechanical insufflation-exsufflation has been increasingly used in individuals with neuromuscular weakness, despite the lack of large randomized trials, although there is cumulative evidence that mechanical insufflation-exsufflation increases cough peak flow and, in combination with noninvasive ventilation, can prolong survival. The literature varies regarding appropriate settings of inspiratory and expiratory pressures, which range from 20 to 60 cm H_2O in positive and negative pressures. Noninvasive ventilation and mechanical insufflation-exsufflation are reported to be less effective in patients with bulbar involvement, and those with spastic upper motor neuron weakness are least likely to tolerate mechanical insufflation-exsufflation because upper airway collapse and/or spasm may be generated. In particular, gradual development of bulbar dysfunction may influence the extent to which a successful use of mechanical insufflation-exsufflation is to be expected.[4]

Third, in patients with bulbar symptoms, noninvasive ventilation and mechanical insufflation-exsufflation should be managed individually, with the expectation that settings and interventions may need to be modified with disease progression. In patients with ALS, the clinical response to noninvasive use of mechanical insufflation-exsufflation is variable, which indicates a certain motor dysfunction of the upper airways. In some cases, the effect of mechanical insufflation-exsufflation seems to be reduced in parallel to disease progression and deteriorating bulbar dysfunction. It is difficult to predict beforehand which individuals will succeed because there are no evidence-based predictive factors.

Treatment failure with mechanical insufflation-exsufflation in patients with bulbar symptoms is likely to be caused primarily by laryngeal adduction during insufflation, predominantly at the supraglottic level. This response precludes air filling of the lungs during insufflation, which causes discomfort and subsequent inefficient exsufflation. A similar approach is appropriate in starting noninvasive ventilation; high inspiratory pressures may generate airway closure, and so pressure should be gently titrated upward.[5]

The main methodological problem in the study was the gradual increase in pressure during the intervention to familiarize the subjects to the feeling of mechanical insufflation-exsufflation. This could be a confounding factor; the subjects become familiarized with the feeling of mechanical insufflation-exsufflation through experience and modification of movements according to the theory of motor learning. Randomizing the intervention sequences could prevent this issue, but, in clinical settings, pressures are always increased gradually to be as gentle as possible for patients.

Individually customized settings for pressure and flow can improve and extend the use of noninvasive mechanical insufflation-exsufflation in ALS, and flexible laryngoscopy can be an efficient tool in selected patients who do not respond as expected. When we increase our understanding of these issues, we can, it is hoped, develop better clinical instructions, more fine-tuned settings, or even new functions for mechanical insufflation-exsufflation devices.

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Laryngoscopy Can Be a Valuable Tool for Unexpected Therapeutic Response in Noninvasive Respiratory Interventions

In reply:

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