Go With the Flow: Are We Cracking the Code? Respiratory Management of Bulbar ALS Is Evolving

Health professionals tend to work in silos. As for the respiratory field, otolaryngologists engage in the upper airways, pulmonologists in the lower airways, and technical devices are applied to treat conditions either in the upper or the lower airways. The larynx is an organ placed centrally in the airways in between these 2 divisions, serving highly specialized tasks vital to normal function.[1,2] Why should all in the respiratory field consider the larynx? The answer is there is mounting evidence that the larynx is strongly involved in the pathophysiology of an increasing number of airway symptoms and diseases, and its function is vulnerable to a variety of internal and external influences.

Amyotrophic lateral sclerosis (ALS) is a motor neuron disease with a complex genetic base, where the clinical picture relates to *both* upper and lower motor neuron degeneration. These neurons control voluntary muscle. Laryngeal dysfunction may be caused by lower motor neuron failure of abductor muscle and/or upper motor neuron hyper-reactivity of adductor muscle.[3] The use of mechanical insufflation-exsufflation (MI-E) to aid secretion clearance has become routine in the respiratory care of patients with ALS, and a topic of increasing interest. An effective cough is related to a deep breath or insufflation followed by an increase in the expiratory air flow to move the mucus proximally.[4] Whereas MI-E treatment may be straightforward in patients with non-bulbar ALS, the treatment is more challenging in patients with bulbar muscle dysfunction.[5-8] In this issue of Respiratory Care, Sancho and colleagues[9] analyzed flow and pressure graphs generated by MI-E treatment to detect unwanted upper-airway responses and state this could aid in selecting the optimal treatment parameters in patients with ALS. Airflow obstruction during insufflation was stated to relate to predominant bulbar upper motor neuron dysfunction, whereas airflow obstruction during exsufflation mainly was due to bulbar lower motor neuron dysfunction.[9] Their work is an important contribution to the field of laryngeal involvement in respiratory care, hereby including flow curve assessments to evaluate the complete cycle of assisted cough. The same group was pioneers with visualization of upper airways during MI-E treatment. In 2004, they examined the upper airways of 3 patients with ALS by computer tomography at baseline and during the exsufflation phase of MI-E treatment, revealing dynamic upper-airway collapse during exsufflation.[8] However, scans were not performed during the insufflation phase at this point. Since then, studies applying direct laryngeal views by transnasal fiberoptic laryngoscopy (TFL) have indicated that MI-E treatment cause adduction of laryngeal structures during insufflation in groups of patients with ALS with progressive bulbar palsy and pseudobulbar palsy. Laryngeal adduction at insufflation precludes free air flow and air filling of the lungs, thereby compromising MI-E treatment.[5,6]

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When analyzing cough effectiveness, the whole cough cycle is important. Sancho et al[9] not only considered the cough peak flow (CPF) but the whole air flow graph and calculated cough volumes during the MI-E cycles. We believe this is an important step in the right direction. The limitations of using CPF as an outcome measure for MI-E treatment in bulbar patients have been highlighted repeatedly,[7,10-12] and particularly the study by Lacombe et al[11] which revealed an abrupt flattening of the exsufflation flow curve in the presence of an otherwise increasing CPF. They, therefore, suggested that using CPF alone failed to detect upper-airway collapse during MI-E and that studying the complete exsufflation air flow curve could reveal laryngeal opening or closure.[11] Taken together, Sancho[9] and Lacombe et al[11] findings indicate that CPF measurement alone may provide a false reading of MI-E effectiveness, whereas the analysis of flow-time graphs helps adjust the parameters.

Sancho et al[9] concluded that visual analysis of air flow graphics may be used to select the optimal parameters during mechanically assisted cough. Similarly, TFL has appeared a reasonable approach in patients who do not respond to MI-E treatment as expected.[5-7] Probably neither of these assessments can yet provide the whole picture of

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the laryngeal air flow due to complex processes categorized as "fluid (or gas)-structure interactions."[13] As the air flow through a tube exceeds a critical velocity, the flow becomes more turbulent; thus, the pressure drops and the air flow causes deformations within the tube (structure), both wanted and unwanted.[13] Wanted deformations are the coordination of glottic closure and opening to perform cough, which affect the air flow, read as the CPF. Unwanted deformations appear when the air flow evokes the fine-tuned laryngeal motor responses and reflexes,[1,2,5,14] resulting in constriction and thus obstruction. Visual laryngeal observation may provide important clinical information in addition to visual analysis of air flow graphics, as a direct view will provide the sequence of events leading to air flow obstruction. An example is a retroflex epiglottis leading to swallowing and flow abruption in a patient; in such a case, the patient should be guided to perform only one or few MI-E cycles instead of longer series.[6] Ultimately, flow and pressure measurements should be added to the setup with TFL.[7] Sancho et al's assumptions that the TFL could interfere with the flows generated during MI-E sessions, whereas use of local anesthesia could interfere with upper-airway reflexes,[9] are aspects for future research.

So what do we know so far? In many patients with ALS, MI-E is applicable, effective, and provides great clinical benefits.[4] As stated earlier by Simonds,[15] ultimately the patients with bulbar ALS have not failed MI-E or noninvasive ventilation—the therapy/therapists may have failed them. Clinicians need to understand both the pathology and to provide careful titration of MI-E parameters to make a difference. Neurologists should be included when subcategorizing patients in a complex progressive disease as ALS. Adduction of supraglottic structures limits insufflation in patients with bulbar ALS[5]; the same occurs during ALS disease progression, where the first signs of laryngeal adduction occur with the highest insufflation pressures and prior to any clinically evident signs of bulbar involvement.[6] Sancho et al[9] included larger numbers of subjects, again indicating that predominant upper or lower motor neuron affection may have different challenges. There is an agreement that individually customized settings seem to promote more optimal response to MI-E treatment.[4-6,9,10] The therapists must use active troubleshooting when treatment does not provide instant effect. Both the analysis of the air flow graphs and in some cases additional TFL assessment might be necessary to characterize adverse responses throughout MI-E interventions. Great care must be taken to avoid applying pressures and cough cycles that the patient's larynx is unable to handle. Further, it is sensible that the terminology used to express both the disease characteristics and upper-airways responses should be both considerate and accurate.[15]

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