Continuous Mouthpiece Daytime Amyotrophic Lateral Sclerosis in Noninvasive Ventilation: Definitive Solid Therapy?

To the Editor:

Treatment options for amyotrophic lateral sclerosis (ALS) are very limited.[1] In some cases, noninvasive ventilation (NIV) by mouthpiece ventilation is a technique that is useful in the treatment of chronic ventilatory failure, particularly in neuromuscular patients.[2] The literature on mouthpiece ventilation in ALS is limited, and there are no published evidence-based guidelines concerning mouthpiece ventilation. Its application is mainly based on the experience of a few centers. Mouthpiece ventilation is particularly useful when there are problems with conventional masks. Patient selection for adequate bulbar muscle function is of paramount importance, and in these patients, mouthpiece ventilation was shown to be an effective alternative to tracheostomy, optimizing convenience and communication.

We have read with great interest the study by Bédard and McKim[3] that concludes that mouthpiece ventilation is a safe, convenient, and effective way to provide NIV in ALS with adequate bulbar function requiring continuous ventilatory support. Although this study shows an original contribution for NIV in severe ALS, we believe that there are some key issues that require evaluation.

First, the addition of mouthpiece ventilation was based primarily on the objective of improving quality of life related to prolonged hours of inconvenient and uncomfortable mask use (>12 h daily) (24 of 37 subjects) and symptoms of dyspnea (9 of 37 subjects).[3] We believe other aspects need to be considered.

Second, the evaluation of bulbar function remains subjective as a clinical assessment, where some clinical tests (swallowing and speech) and patient recall define the level of bulbar involvement. This is in contrast to the evaluation of ventilation, where several objective functional markers are followed through time. Of all of the routine respiratory tests ordered, cough peak flow is the only test that reflects bulbar function in addition to inspiratory and expiratory muscle function. The difference between cough peak flow and peak expiratory flow has thus been proposed to measure bulbar involvement.[4]

Third, the bulbar subscores of the Revised Amyotrophic Lateral Sclerosis Functional Rating Scale discriminated between those subjects having sufficient bulbar function to succeed with mouthpiece ventilation and those who did not. It seems to be a simple and useful tool to assess candidacy for mouthpiece ventilation. In this study, the scores' cutoff points are arbitrary, and no study has defined a cutoff that accurately reflects an objectively measured clinical impairment. We need a more accurate way to detect and quantify the level of bulbar involvement.[4] For example, a specific test of bulbar function may help to recognize the group of patients at higher risk of NIV failure or in need of early initiation of mechanical cough assistance, or it may simply help to better classify/quantify bulbar involvement[4] Further prospective clinical trials to confirm the efficacy of mouthpiece ventilation should be offered as a rational alternative to tracheostomy for individuals able to hold a mouthpiece, particularly in individuals with non-bulbar ALS needing continuous ventilatory support.

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Continuous Mouthpiece Daytime Amyotrophic Lateral Sclerosis in Noninvasive Ventilation: Definitive Solid Therapy?—Reply

In reply:

We appreciate very much the opportunity to respond to the letter from Drs Fiorentino and Esquinas with regard to our publication on mouthpiece ventilation in amyotrophic lateral sclerosis (ALS).[1] We are pleased that it was read with great interest, and we sincerely thank our subjects for allowing us the opportunity to document the effectiveness of mouthpiece ventilation. Providing ventilator support and airway clearance to individuals with ALS is often challenging; therefore, developing more accurate means of assessment and novel techniques of support, such as mouthpiece ventilation, is critical, particularly in avoiding unnecessary tracheostomy placement.

We are in full agreement with many of the statements expressed in this letter and recognize the importance of mutual effort in establishing evidence-based and best practices in the respiratory care of those with ALS. Sadly, even in the Canadian Thoracic Society clinical practice guideline on home mechanical ventilation,[2] there are no specific recommendations for mouthpiece ventilation for patients with ALS, although there are for patients with Duchenne muscular dystrophy. The recommendations for up to 24-h noninvasive ventilation (NIV), rather than tracheostomy, although not explicit, could be interpreted to include mouthpiece ventilation, clearly a form of NIV. We also agree that the paper demonstrates that mouthpiece ventilation is safe, convenient, and effective and that, indeed, key issues are still in need of further evaluation.

In particular, we agree entirely that a more objective assessment of bulbar function and its potentially predictive relationship to success with mouthpiece ventilation would be helpful rather than relying on the first 3 questions of the Revised Amyotrophic Lateral Sclerosis Functional Rating Scale score, referred to as the bulbar subscore (b-ALSFRS-R). Within individuals, this score may vary from time to time, and in our experience, it may differ significantly from a more objective clinician assessment. However, we do not have consistent access to more objective measures of bulbar function and must rely on this value as a subjective estimate. It may also be of greater utility in studying populations of ALS patients as opposed to single individuals. Not even we, however, would rely entirely upon