

# Non-Invasive Ventilation for Canadian Patients with ALS: Do We Have Consensus?

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Initiation of non-invasive ventilation (NIV) plays an important role in the management of patients with Amyotrophic Lateral Sclerosis (ALS). Class I evidence exists from a randomized controlled trial demonstrating a median survival benefit of 205 days in ALS subjects treated with NIV<sup>1</sup>. Studies have also demonstrated that NIV enhanced the quality of life of patients with ALS resulting in an improvement in sleep quality, energy levels, symptoms of shortness of breath, and daytime somnolence, along with a decrease in episodes of depression<sup>2-4</sup>. Non-invasive ventilation should be accessible to all patients with ALS in Canada and this is reinforced in the recently updated American Academy of Neurology Practice Parameters that suggests NIV be used to treat respiratory symptoms and to prolong survival<sup>5</sup>.

In this issue of the Canadian Journal of Neurological Sciences, Ritsma et al report the results of a survey completed by clinicians from 11 of the 15 major academic ALS clinics across Canada<sup>6</sup>. The authors inquired about each clinic's practice regarding the number of ALS patients receiving NIV and invasive ventilation, the markers and thresholds for initiating NIV and what potential barriers exist in patients receiving NIV. They found that Canadian clinicians deem respiratory symptoms (orthopnea, dyspnea, and morning headaches likely secondary to CO<sub>2</sub> retention) as the most important markers in determining when to initiate NIV. Additional testing such as nocturnal oximetry, forced vital capacity (FVC) and morning blood gas were also reported by respondents to guide their decision making.

The management strategy reported by the Canadian ALS clinicians is validated by the latest American Academy of Neurology Practice Parameters<sup>5</sup>. However, in addition to routine monitoring of respiratory symptoms, best evidence suggests that nocturnal oximetry and maximal inspiratory pressure are the most sensitive respiratory monitoring measures. It has also been demonstrated that supine FVC is likely more sensitive than upright FVC in detecting diaphragmatic weakness.

An important finding of the survey conducted by Ristma et al<sup>6</sup> relates to ongoing barriers for ALS patients to receive NIV and the perception by a number of respondents that this important intervention may be underutilized. The most frequently perceived barriers were secondary to patient intolerance of the NIV apparatus and lack of accessibility to a respirologist and ventilation technologist. These barriers should be explored further in individual clinics to confirm that experienced ventilation technologists are fitting patients for the NIV masks which can often be adjusted and modified to improve comfort and compliance. Given the limited number of multidisciplinary ALS clinics across Canada, institutions must

adequately equip and support their clinics to provide timely patient access to respiratory professionals and equipment when necessary. Appropriate support and infrastructure in multi-disciplinary ALS clinics is essential to maintaining a high standard of care and is ultimately 'cost-saving' as patients without timely access to NIV will likely end up in the emergency rooms and admitted to hospital. With consensus guidelines and recommendations on wait times for ALS patients to receive interventions such as NIV and gastrostomy tubes, clinics can determine if their delivery of care falls below the standard and petition to obtain additional resources from their institutions.

The study is limited by the small number of respondents and recall bias that is inherent to survey research. In addition, there is an ascertainment bias which may affect the generalizability of the results as surveys were not returned from a significant portion of ALS clinics that may have very different practices. However, this study illustrates that variability does exist in ALS centres across Canada regarding the management of respiratory symptoms and highlights the importance of obtaining consensus criteria. Although care must be individualized, which is of great importance for a heterogeneous disease like ALS, practice parameters based on best available evidence are essential to help guide clinicians to provide the highest quality of care.

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