


Long term survival in motor neuron disease. Insights about noninvasive ventilation efficacy

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To Editor

We have read, with great interest, this study by Walsh et al.¹ where an extensive population of patients with NMD was analyzed.

The study is a retrospective and is very extensive and it is a good snapshot of the use of positive pressure noninvasive mechanical ventilation in this population.

There is now evidence that the early start of a respiratory monitoring program of neuromuscular patients allows us to identify the best time to start therapy with a non-invasive assisted ventilation, which results in an improvement in the quality of life with an increase in survival.^{2–5}

The Authors described a longer survival with the use of NIV. However, some aspects of the clinical course were unknown during this long period of observation (5-year period). We noted that hospital admissions due to an exacerbation of Motor neuron disease or other causes of decompensation during observation period as a cause of mortality were not analyzed in this study. Furthermore, the progression of the disease and nutritional status and the possible improvement by applying appropriate nutrition measures was not assessed.

We consider that this data would impact survival and must be considered. It was also not indicated how many patients were subjected to NIV due to waking respiratory failure, and how many due to the presence of respiratory failure only during sleep, as well as the type of interface used, which, in our opinion, can have a clear impact on NIV efficacy and compliance result. There are also no data on the blood gas analysis at the beginning (pre-NIV) and during follow-up, which could have been predictive on the effectiveness of NIV and the patient survival (together with the

measurement of the strength indices of the respiratory muscles, which were reported at zero time), as well as data on the NIV setting were reported only at baseline, but we do not know if these parameters have been adjusted and which way during the observation period, if this directly correlated to the progressive worsening of respiratory function, nor if it has been related to mortality.

With compliance to NIV, it has been described that those with bulbar MND had better compliance and machine utilization, poorer respiratory function had a greater need for NIV than the non-bulbar group; then, it was observed that in general if a patient was tolerant of the machine within 3 months of initiation, he remained compliant throughout the rest of the time of observation. These aspects need further clarification by the authors, as there is little information on these associations during the observation period. The authors did not evaluate other tools in non-compliant patients such as positive abdominal pressure ventilation or

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mouthpiece ventilation which, especially in the initial stages of the disease, could guarantee good compliance. Evaluation of the data for which the patients did not start mechanical ventilation (such as non-compliant family environment, etc.) or of any side effects or decubitus would also have been useful in the study. Moreover, it is perhaps the first study carried out in a patient cohort in the south of Ireland, and this leaves further room for reflection on the impact of the cultural background on compliance with the use of NIV in the long term.

It would also have been interesting to have data on the number of patients who were then possibly subjected to tracheotomy, and for how many months NIV was used before the occurrence of this procedure, as a further confirmation of the effectiveness of the efficacy of early and long-term NIV, in addition to survival data

Apart from these statements, the results of this work are still in line with what is present in the literature of the last 15 years,^{2-4,6} about the effectiveness and importance of the early use of NIV, in ALS patients.^{2,6} We also believe that the results and conclusions of this study, require some clarification.

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References

1. Walsh LJ, Deasy KF, Gomez F, et al. Use of non-invasive ventilation in motor neuron disease - a retrospective cohort analysis. *Chronic Respir Dis* 2021; 18: 14799731211063886, DOI: [10.1177/14799731211063886](https://doi.org/10.1177/14799731211063886)
2. Bourke SC, Tomlinson M, Williams TL, et al. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurol* 2006; 5: 140–147.
3. Bach JR. Amyotrophic Lateral Sclerosis. *Chest* 2002; 122(1): 92–98.
4. Radunovic A, Annane D, Rafiq MK, et al. Mechanical ventilation for amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev* 2017; 2018: CD004427. DOI: [10.1002/14651858.CD004427.pub4](https://doi.org/10.1002/14651858.CD004427.pub4)
5. Moretti M, O’Sullivan E, Eustace J, et al. Il percorso respiratorio dei pazienti con sclerosi laterale amiotrofica. Gruppo di Lavoro Pneumologico dei Presidi Sanitari accreditati dalla Regione Emilia Romagna per il trattamento dei pazienti affetti da sclerosi laterale amiotrofica. *Rassegna di Patologia dell’Apparato Respiratorio* 2007; 22: 391–406.
6. Carratù P, Spicuzza L, Cassano A, et al. Early treatment with noninvasive positive pressure ventilation prolongs survival in Amyotrophic Lateral Sclerosis patients with nocturnal respiratory insufficiency. *Orphanet J Rare Dis* 2009; 4: 10, DOI: [10.1186/1750-1172-4-10](https://doi.org/10.1186/1750-1172-4-10)