The benefit of non-invasive ventilation in motor neuron disease. Response to letter CRD-22-0013

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Motor neuron disease, non-invasive ventilation, respiratory failure, survival, pulmonary function tests

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Dear Editor,

We would like to thank Dr Fiorentino and his colleagues for their interest in our publication and for the opportunity to clarify the issues which they have highlighted.

Our study was a retrospective cohort analysis of motor neuron disease (MND) patients attending a service in the South of Ireland. The focus of the study was to determine whether or not we could establish a relationship between patient survival and non-invasive ventilation (NIV) usage in this cohort. We acknowledge that due to its observational and retrospective nature direct causation is more difficult to establish and certainly we agree that there may be a variety of factors contributing to the outcome observed. Despite these limitations we believe that the results and observations of this study are nevertheless valid.

The preferred approach to NIV set-up in our institution is to set patients up and establish compliance where possible as early as possible. Hence, the majority of patients underwent elective set up either at home or as an inpatient rather than during an acute deterioration. The analysis of acute hospital admissions and how this may have affected respiratory functioning was beyond the scope of this study as was an evaluation of why patients may not have been compliant. The aetiology of non-compliance is multifactorial, and given the nature of this study, the vast majority of data was collected retrospectively. Unfortunately, given the natural history of MND, at the time of data analysis the majority of our patient cohort were not alive and so even prospective collection of this data would not have been feasible.

In our institute we do not routinely perform Arterial Blood Gas (ABG) sampling once NIV usage has been established. Patients undergo assessment to commence NIV, only after the diagnosis of MND has been clearly established. Patients are usually admitted and undergo clinical assessment, ABG, overnight pulse oximetry and pulmonary function testing. It is known that the majority of patients with MND have evidence of respiratory muscle dysfunction at the time of diagnosis.1,2 As Dr Fiorentino and colleagues stated there is now a known benefit to early commencement of NIV. Therefore, if patients were willing to attend, they were commenced on NIV as early post diagnosis as possible, usually using a full mask interface. Titration of NIV settings wasn’t recorded during this study.

We do comment on the measurement of pulmonary function testing with disease progression, but we note in our limitations section that this data set was incomplete and only available for 78 patients.

We used the observation that if a patient was compliant at 3 months, they would remain compliant in order to simply the fact that we had low numbers in groups. We acknowledge again that compliance is variable and can vary over hours, days, and weeks. Indeed, we also mention that compliance data was not always available or if it was may not have been regularly recorded which reduced available data.

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We acknowledge that PEG feeding may also contribute to improved survival and indeed found that there was a slight difference in the total group which seemed to favour PEG insertion. We did not however find a difference in survival in those with bulbar MND and PEG insertion (20.2 months) compared with survival in bulbar MND without PEG (19.15 months). It should be noted that only 27.8% of our cohort however were PEG fed. Tracheostomy is not favoured in Ireland.

Again, it should be noted that this was a retrospective analysis of a service. Changes occurred within the service during the time of the observation period such as improvements in staffing numbers and in recording of vital data such as pulmonary function testing results and compliance data. This is acknowledged in the original manuscript. We would suggest that a repeat, smaller, prospective study may be beneficial to validate the results observed. The overall conclusion of our study was that NIV has a useful role to play in the management of patients with MND.

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