Motor neurone disease in the greater Wellington region: an observational study

Viswas Dayal, Ian Rosemergy, Janet Turnbull

Abstract

Aims: This study investigated the demographic and disease characteristics of motor neurone disease (MND), as well as specific aspects of healthcare provision for MND patients in the greater Wellington region.

Methods: Data was collected from clinical records of all known patients with a diagnosis of Motor Neurone Disease MND in the Capital and Coast and Hutt Valley district health board catchment zones over a 12-month period. Survival data was collected at 2-year follow-up.

Results: The study population consisted of 40 patients. The mean age at diagnosis was 66.2 years (SD of 13.2). Thirty patients (75%) were New Zealand European. Predominantly limb signs were present in 12 (30%), and 11 (27.5%) had bulbar signs, while 14 (35%) had a mixed pattern and a further 3 (7.5%) had respiratory muscle weakness. At 2-year follow-up, the median survival time following symptom onset in the 32 deceased patients was 29 months (range 6–126 months).

Conclusions: The demographics and disease characteristics of MND in this cohort of patients from the Wellington region is similar to those found in overseas studies. The majority of patients received care in accordance with established guidelines, although certain aspects of healthcare delivery could be further improved.
Method

This was an observational and descriptive study using data from clinical records. The district health board protocol for ethics approval was followed, and no ethics committee approval was required for this study given it consisted of collecting observational information in the form of an audit, with no patient contact or interventions carried out.

Patients with a diagnosis MND were identified using hospital clinical coding data at Hutt Valley Health and Capital and Coast District Health Boards, which included presentations to Wellington Regional, Hutt and Kenepuru Hospitals. We reviewed all outpatient visits as well as inpatient encounters between 1 June 2011 and 31 May 2012.

In addition to this, the MND field workers for the Wellington and Hutt Valley regions were contacted to provide registries of all known patients with MND during this 12-month period, and any additional patients not identified via clinical coding information were added to the study group. No patients or families were contacted as part of the study.

The following data was collected: Age at diagnosis, sex, ethnicity, source of referral, clinical manifestation of MND at presentation (i.e. predominantly bulbar, limb or respiratory involvement), MND subtype, family history of MND, presence of cognitive impairment, and use of the disease-modifying drug riluzole (Rilutek).

Survival data was collected at 24 months following the end of the study period, in June 2014. This was done by calculating the time from onset of symptoms to death for each deceased patient. Aspects of community health care provision for MND patients that were evaluated included the number of patients who received respiratory assessment and non-invasive ventilation, percutaneous endoscopic gastrostomy tube placement, speech language therapist and dietitian input, and palliative care input.

Results

A total of 40 patients were identified as having a diagnosis of MND during the study period in the greater Wellington region. Twenty-eight of these were from the Capital and Coast District Health Board catchment zone and 12 from Hutt Valley Health.

The mean age at diagnosis was 66.2 years (SD of 13.2) and the median was 67 years, with a range of 27–94 years. There were 22 males and 18 females. The 12-month prevalence of MND in the Wellington region based on this data and the most recent census is 8.5 cases per 100,000.

The breakdown on subtype of MND was as follows: 35 patients had amyotrophic lateral sclerosis (ALS), three patients had primary lateral sclerosis (PLS), one had progressive muscular atrophy and one had progressive bulbar palsy.
Table 1. Demographic and clinical characteristics of MND patients in the Wellington region

<table>
<thead>
<tr>
<th>Age interval (years)</th>
<th>N (%)</th>
<th>Ethnicity</th>
<th>N (%)</th>
<th>Clinical manifestation</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>21–30</td>
<td>1 (2.5)</td>
<td>NZ European</td>
<td>30 (75)</td>
<td>Limb involvement</td>
<td>12 (30)</td>
</tr>
<tr>
<td>31–40</td>
<td>2 (5)</td>
<td>Maori</td>
<td>2 (5)</td>
<td>Bulbar involvement</td>
<td>11 (27.5)</td>
</tr>
<tr>
<td>41–50</td>
<td>1 (2.5)</td>
<td>Asian</td>
<td>2 (5)</td>
<td>Mixed limb and bulbar</td>
<td>14 (35)</td>
</tr>
<tr>
<td>51–60</td>
<td>5 (12.5)</td>
<td>Pacific Islander</td>
<td>1 (2.5)</td>
<td>Limb with respiratory involvement</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>61–70</td>
<td>20 (50)</td>
<td>Other European</td>
<td>5 (12.5)</td>
<td>Bulbar with respiratory involvement</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>71–80</td>
<td>6 (15)</td>
<td></td>
<td></td>
<td>Isolated respiratory involvement</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>81–90</td>
<td>4 (10)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>91–100</td>
<td>1 (2.5)</td>
<td></td>
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</tbody>
</table>
The referral sources to neurology in this group of patients were from the patient’s general practitioner 40% (16/40), hospital physician 27% (11/40), other hospital team 8% (3/40) and unknown 25% (10/40).

Hospital physician referrals consisted of six from general Medicine, four from geriatric services, and one from rheumatology. Other hospital services referring were orthopaedics (two) and ENT (one). Of the unknown group, some patients were transferred to the public hospital service from private Neurology consultations and did not have any referral on record.

Fronto-temporal dementia was diagnosed in one of the 40 patients. Mild cognitive impairment was documented in a further three. Emotional lability with a pseudobulbar affect was noted to be present in 7 patients, with 5 of these having bulbar predominant disease.

A positive family history of MND was noted in 22% (4 of 18 patients) in whom a documented enquiry was found. This is higher than expected but is likely to represent a reporting bias in the subgroup.

Only 2 of the 40 patients in the cohort were on the medication riluzole. With review of the survival data at 24 months, we found that 32 patients in the cohort were deceased and the median survival from the time of symptom onset was 29 months (range 6-126 months). The mean survival time was 37.5 months (SD of 29.1).

Twenty-one patients received a Respiratory physician assessment with spirometry. A further six had spirometry only. Ten patients (25%) in total received non-invasive ventilation with Bi-level positive airway pressure (Bipap).

Dietitian input was recorded in 21 patients and speech language therapist (SLT) input was recorded in 27 of the 40 patients. All except three patients with bulbar disease were assessed by an SLT. A PEG tube was placed in 12 patients out of 26 with bulbar disease. A discussion regarding consideration of a PEG was documented in three further patients, two of whom declined the procedure and one died prior to tube placement. One patient with bulbar predominant disease received salivary gland Botulinum toxin injections for excessive salivation.

Palliative care services are provided by Mary Potter Hospice in the Capital and Coast region and Te Omanga Hospice in the Hutt Valley region. Twenty-four patients in total were on the respective hospice programmes, and documentation of a discussion regarding future referral was made in a further three.

**Discussion**

Our findings show that the peak prevalence of MND in this population is in the seventh decade with a male to female ratio of 1:1.2. This is consistent with data from other epidemiological studies overseas.1–3,9,13

We found a 12-month prevalence of 8.5 per 100,000 in the Wellington region, and although this is slightly higher than reported figures of 5-8 per 100,000 elsewhere, the point prevalences in these studies may not be directly comparable. Despite this, there is a suggestion from another study that the incidence rates of MND in New Zealand is high relative to overseas populations, and has been increasing over the last few decades.

The majority of patients had the ALS form of MND in this group and 35% presented with mixed limb and bulbar symptoms. Overall, 65% of patients had some bulbar symptoms at presentation. Bulbar presentation is associated with a worse prognosis than spinal onset disease and early identification of this patient subgroup is important in planning subsequent management.
Respiratory involvement in isolation was uncommon at 2.5%. Other studies looking at the first clinical manifestation of MND report bulbar onset in 15-25%, 9,11,13 respiratory muscle weakness as the first manifestation in less than 3% 12 and spinal or limb onset in the remainder, although one large study of MND phenotypes reported approximately equal incidences of classic and bulbar types, 14 as we found in this cohort.

Our data also confirms that general practitioners were the single largest group of doctors who had first contact with MND patients and made the most referrals to the neurology service for a specialist opinion.

The median survival time of 29 months (2.4 years) for the 32 of the deceased patients on collection of data at 24 months may be influenced by a selection bias of excluding patients with relatively slowly progressive disease who tend to survive longer and did not have survival data available as they were still living at this follow up period. Nevertheless, this figure is similar to reported longevity in MND patients in larger studies, 9,10 including one that reported data from a local Canterbury cohort. 20

Treatment options for MND are limited and riluzole is the only drug available that has any impact on survival, albeit with a modest effect of prolonging survival by 2–3 months. 15 Riluzole has recently been approved for funding for MND patients by PHARMAC upon application via special authority, but was not funded during the period of the study and had a cost to the patient of $700 per month. 17 This may explain the low utilisation rate in this cohort. It is possible that more MND patients would now opt to take the drug given it is subsidised.

The American Academy of Neurology (AAN) guidelines 16 recommends respiratory function tests at diagnosis for all MND patients. Almost seventy percent of patients in this cohort had spirometry assessments early after their initial diagnosis. This could be improved with the goal of aiming to objectively assess respiratory function in the remaining patents. However, some of these patients had significant bulbar dysfunction which would preclude them from the use of non-invasive ventilation and may explain why respiratory function tests were not performed.

A PEG tube placement or a documented discussion about future insertion was carried out in over half of patients in the cohort with bulbar dysfunction. The AAN guidelines recommend that a PEG be offered to all patients with symptomatic dysphagia, and that the optimal efficacy and safety of the procedure is when the vital capacity on spirometry is greater than 50% predicted.

Overall, while there remains some room for improvement, it appears that the hospital and community MND services within the Wellington and Hutt Valley regions are managing to provide a comprehensive service when compared to international guidelines, for these patients who have a very challenging set of physical and emotional problems.

Competing interests: Nil.

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**References**


