

# A retrospective review of specialist palliative care involvement in motor neurone disease

## Abstract:

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## Abstract

The provision of specialist palliative care to Irish patients suffering from motor neurone disease has not been described in the literature. The purpose of this study was to characterize the care provided at a Dublin hospice. Consecutive referrals between 1st January 1999 and 31st December 2008 (n=72) were reviewed. At the time of data collection, 61 (84.7%) were deceased, 9 (12.5%) were alive and the status of 2 (2.7%) was unknown. At first assessment, 48 (66%) had bulbar symptoms and 35 (49%) had respiratory symptoms, 50 (70%) were receiving Riluzole, 25 (35%) had a feeding tube and 13 (18%) were using non-invasive positive pressure ventilation (NIPPV). Median survival from the point of referral was 7 months (95% CI 4.5 - 9.4). Of the 61 deceased patients, 22 (36%) died at home, 22 (36%) died in the inpatient unit, 9 (15%) died in hospital and 8 (13%) died in a nursing home.

## Introduction

Motor neurone disease/amyotrophic lateral sclerosis (MND/ALS) is a neurodegenerative disease characterised by loss of motor neurons in the spinal cord, brainstem and motor cortex. It has a reported population incidence in Ireland of 2.1 per 100,000<sup>1</sup> and is associated with a median survival of 16.4 months<sup>2</sup>. Though it is relatively rare, its population-impact is significant; it is estimated<sup>3</sup> that for every person diagnosed with MND, another 14 family members or close friends will be impacted<sup>4,5</sup>. The palliative care needs of MND patients have been well documented and explored in the literature<sup>6</sup>. Despite this, specialist palliative care services for MND patients do not appear to be as well established as those for cancer patients. In practice, there appears to be little consistency in the way in which specialist palliative care is delivered within the overall context of MND multidisciplinary care<sup>7,8</sup>. To date, there has been limited characterisation of existing models of MND specialist palliative care<sup>9</sup>, with no data available for an Irish setting. The absence of such data prompted us to undertake a retrospective evaluation of a decade of MND service delivery at Our Lady's Hospice & Care Services, Dublin. This study specifically aimed: 1) to describe the demographic and clinical characteristics of the referred MND population 2) to collect key peri-mortem data and 3) to understand how MND patients used the service.

## Methods

A retrospective review of clinical notes was undertaken. This was conducted at the two hospice units of Our Lady's Hospice and Care Services. Together, these two units serve the specialist palliative care needs of two geographical regions within Dublin. They consist of a home-care service, an inpatient service (one 36-bedded unit and one 12-bedded unit at two different sites), a day hospice service and a consultant-led outpatient service. All patients with a diagnosis of MND that had their first specialist palliative care assessment between 1st January 1999 and 31st December 2008 were identified through the hospice database system. The clinical notes of these patients were obtained for review. Data chosen for collection included demographic data, clinical data relating to early assessment and the last week of life, and service-use data. Permission was obtained from the Ethics and Medical Research Committee at St. Vincent's Healthcare Group Limited prior to data collection. Data were analyzed using SPSS Version 17.

## Results

Over the ten year period there were 73 first palliative care service assessments of patients with a documented diagnosis of MND. The clinical notes of 72 out of the 73 patients were reviewed, as one record was incomplete. At the time of data collection, 61 patients (84.7%) were deceased, 9 patients (12.5%) were alive and the status of 2 (2.7%) was unknown.

!Percutaneous Endoscopic Gastrostomy Non-invasive positive pressure ventilation \*Riluzole is a disease-modifying drug licensed for ALS

### Demographic and clinical characteristics at first assessment

The demographics of the referred population, along with the referral sources, are outlined in table 1. The clinical features are outlined in table 2.

### Time to referral from diagnosis and time to death from referral

Time to referral from diagnosis was available for 71 out of 72 patients. Patients had a documented diagnosis of MND at a median of 14 months (range: 1-238) prior to specialist palliative care referral. Median survival from the point of specialist palliative care involvement was estimated to be 7 months (95% CI 4.5 - 9.4) using a Kaplan Meier survival curve.

Figure 1

### Peri-mortem Data

Place of death is illustrated in Figure 1. Detailed peri-mortem data were only available for the patients who died either in the inpatient palliative care unit or at home under our specialist palliative care homecare team, and analysis was therefore confined to these patients (n=42). Treatments used in the last week of life are summarised in table 3. For those patients who received regular opioids in the last 24 hours of life, the median regular 24-hour oral morphine equivalent dose used was 60mg (range: 10-720mg) and the mean was 116mg (SD, -174.5). For those patients who received regular subcutaneous midazolam in the last 24 hours of life, the median regular total 24-hour dose used was 20mg (range: 3-100mg) and the mean was 27mg (SD, -20.55).

### Service-Use Data

Analysis of service-use was confined to the 61 deceased patients in order to capture the pattern of service-use from the point of referral through to death. The homecare service was the most used service, with 69% (n=42) of patients receiving homecare for a median of 4 months prior to death. The inpatient palliative care unit was used by 54% (n=33) of patients. Patients who used the respite/symptom-control service used a median of 2 respite admissions per person (range: 1-26). The mean and median duration of respite/symptom-control admissions were 21 and 15 (range: 7-120) days, respectively. The mean and median duration of terminal care admissions were 69 and 63 (range: 2-239) days, respectively.

### Discussion

To the authors' knowledge, this is the first reported study describing specialist palliative care delivery to MND patients in an Irish setting, though Irish hospices have contributed data to multi-centre studies<sup>9</sup>. A number of similar studies have been undertaken in the UK setting. In 1981, Saunders et al<sup>10</sup> retrospectively reviewed their experience in 100 patients. A later study at the same institution by Oâ Brien et al<sup>5</sup> reviewed 124 MND patients over the period 1980 to 1990. Unlike our study, both of these studies primarily described the inpatient MND population. Oliver et al<sup>8</sup> were the first to evaluate MND home-care services. Importantly, these previous studies predate the use of potentially life-prolonging therapies in MND/ALS patients, such as enteral nutrition, riluzole and non-invasive positive pressure ventilation. In Ireland, enteral feeding only became widely used from the 1990s onwards<sup>11</sup>. riluzole became available to all ALS patients following licensing in 1996 and NIPPV was not widely used until 2004<sup>11</sup>. Our study has a number of limitations. Like previous studies, the study design is retrospective, thus limiting the amount and the quality of the data collected. Secondly, the peri-mortem data were limited to those patients who died in the inpatient unit or at home. Finally, reported medication dosages may be underestimated as â as required medicationsâ were not recorded.

The study provides further evidence for the heterogeneous presentation and course of MND, in keeping with the increasing recognition of phenotypic heterogeneity in ALS<sup>12</sup>. The unpredictable course of MND makes it difficult to define the point at which a referral should be made to specialist palliative care. European guidelines indicate that early referral to specialist palliative care is often appropriate<sup>13</sup>. The burden of MND care on specialist palliative care services has long been a source of concern, however. In a New Zealand survey of palliative services offering care to MND patients, concerns were raised about bed-availability for respite care, the high level of resources and time needed, and the possible need for long-term care<sup>14</sup>. Cost of MND care is also a particular source of concern. In an American study comparing the cost of hospice ALS care to hospice lung cancer care, it was shown that the mean per patient cost of ALS care is substantially higher than that of lung cancer care<sup>15</sup>. Prognostically-based hospice-referral criteria have been used in some countries to ensure appropriate referrals. Studies have shown criteria, such as the USA Medicare criteria for MND hospice care, to be too restrictive and specific, however, with many MND patients not meeting the criteria despite having specialist needs<sup>16</sup>. In our institution, we accept all referrals and prioritise them according to need. Our data would support this type of referral policy given that the median duration of hospice-use was estimated to be only 7 months.

The peri-mortem data provides information on important palliative care outcomes in MND. Home death occurred in 36% of referred cases, which is higher than the current national figure for home deaths in Ireland of 25%<sup>17</sup>. This is also higher than the reported figure of 15% for home deaths in a multicentre retrospective hospice study, involving one hospice in Ireland and 5 in the UK, during the period 2007-2009. By contrast, another study, which retrospectively analysed deaths in 50 patients referred to a single UK hospice, reported that home death occurred in 55% of patients<sup>18</sup>. Though we did not collect data about the mode of death, use of a syringe driver to subcutaneously deliver medications in only 76% of deaths suggests that a significant proportion of MND deaths may have occurred suddenly. Oâ Brien et al reported that 40% of inpatient hospice deaths occurred within 12 hours of a sudden deterioration. The relatively low use of a syringe driver at the end of life may also be partially explained by the presence of a gastrostomy feeding tube in a significant proportion of patients<sup>5</sup>.

The study adds to the literature about the use of medications in the last week of life in MND. It suggests that MND patients may require relatively low doses of morphine in the last week of life. Other studies report similar results, though they mainly only report mean rather median dose and some record the mean 24-hour dose over the terminal phase as opposed to specifically over the last 24 hours. Oâ Brien et al reported a lower mean morphine equivalent oral dose of 30mg per 24 hours but this actually reflected morphine-use over a mean duration of 58 days. In a study reviewing MND home-care at one particular hospice, a mean morphine equivalent oral dose of 90mg per 24 hours was used but it was not clear whether this calculation was confined to the last 24 hours of life. A recently published multi-centre MND hospice study reported a mean morphine equivalent oral dose of 80mg per 24 hours in the last 24 hours of life<sup>9</sup>. In terms of service-use, our study indicates that respite and symptom-control admission durations rarely exceed 2 weeks. This provides evidence against a common fear of excessive respite-admission length amongst specialist palliative care providers. A UK study reported the same median respite admission length of 15 days<sup>7</sup>.

Riluzole at the end of life has not previously been investigated. Our study indicates that almost 50% of MND patients continue to receive riluzole in the last week of life at our institution. There is no comparable data in the literature<sup>19</sup>. There is strong evidence that 18 months of treatment with riluzole prolongs survival in ALS by about 3 months<sup>20</sup>. It is not known, however, whether it has any effect in later stage ALS and it is therefore not clear if and when it should be discontinued<sup>20</sup>. The NICE (National Institute for Clinical Excellence) guidance states that, in the latter stages of their disease, patients should be provided with the opportunity to discontinue treatment if the responsible clinician considers it appropriate<sup>20</sup>. Inappropriate continuation of riluzole has financial implications, given that the estimated cost of making riluzole available to all ALS patients in England and Wales is 7.5 million per annum<sup>20</sup>. In conclusion, this study suggests that a flexible model of specialist palliative care service delivery is necessary to accommodate the heterogeneous needs, trajectories and experiences of MND patients. Prospective,

multi-centre studies are needed to further investigate how best to incorporate specialist palliative care into their overall care.

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