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Clinical Study

Activities of daily living in motor neuron disease: role of behavioural and motor changes

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ABSTRACT

Impairment in the activities of daily living (ADL) in motor neuron disease (MND) has been little investigated. The contributions of both behavioural and motor changes on functional performance have not been explored. A postal survey in New South Wales, Australia, included assessments of ADL, behavioural change (carer-based) and MND severity. Eighty-two patients were subdivided into groups according to onset presentation: bulbar ($n = 23$) and limb ($n = 59$). There were significant differences in ADL performance between limb and bulbar onset groups depending on ADL task. Disability was also dependent on disease severity as measured by the Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised (ALSFRS – R) score. Importantly, variance in ADL scores was dependent on both motor and behavioural factors. This study confirms the progressive disabling nature of MND, which is dependent on disease severity and shows qualitative differences depending on onset presentation. A model combining motor and behavioural changes explained 57% of variance on ADL performance, with important implications for clinical intervention.

1. Introduction

Although motor neuron disease (MND) was originally perceived as a pure motor syndrome, recent studies have revealed frontal lobe dysfunction with cognitive and behavioural changes, such as apathy and disinhibition, in up to 50% of patients, with 10% to 20% of patients fulfilling diagnostic criteria for frontotemporal dementia (FTD).[1,2] The presentation and pathology of FTD is heterogeneous but an important subgroup has TDP43-positive pathology, which is identical at a cellular level to that found in MND,[3,4] strengthening the overlap between FTD and MND.

Patients with FTD have considerable impairment in activities of daily living (ADL), which can be more severe than in Alzheimer's disease depending on FTD subtype.[5] In addition, a recent study established that frontal dysfunction (apathy, disinhibition and executive dysfunction) accounted for most of the functional disability in behavioural variant FTD.[6] Given that a proportion of MND patients present with cognitive dysfunction as well as physical impairment, the various contributions of each factor to ADL impairment is clearly an important but unresolved question. It is not clearly established whether cognitive dysfunction in MND is associated with bulbar or limb onset.[1,7,8] Understanding how these factors can interact has clear implications for patients and their families, as well as treating clinicians.

Functional disability is commonly measured using specific scales of ADL. These scales are generally subdivided into basic ADL (BADL) and instrumental ADL (IADL), depending on their complexity. BADL are those related to everyday core tasks such as eating, hygiene, dressing, while instrumental activities comprise more complex ones such as meal preparation, shopping, housework and managing finances.

Surprisingly few studies have focused on ADL disability in MND, and none has yet addressed the underlying behavioural correlates of disability. One study investigated whether general functional assessments were sensitive to disability in MND,[9] while another explored the contribution of functional impairment to patient psychological distress.[10] It remains unknown, however, how functional decline relates to the severity of MND.[11] It may be predicted that upper limb dysfunction is a major determinant of ADL given their dependence on hand function, but the contribution of behavioural changes in functional disability in MND is unknown. As such, the aims of this study were to: (i) describe functional disability in MND in terms of BADL and IADL, taking into consideration disease severity as measured by the Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised (ALSFRS – R) score[12]; (ii) investigate whether limb and bulbar onset presentations yield different levels of ADL performance; and (iii) explore which motor and behavioural factors can explain patient disability in MND.

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2. Materials and methods

2.1. Participants

A postal survey was conducted in New South Wales, Australia, with the support of the MND Association. Dyads of carer–patient members of the association were asked by letter if they were interested in taking part in this survey. Of 122 patients with ALS who accepted an invitation to take part in this survey, 82 dyads of patients-carers (67.2%) responded to the questionnaires, which were mailed by post. All patients had been diagnosed by a neurologist. Ethics approval was obtained from the South Eastern Illawarra Ethics Committee and informed consent was given by all dyads of carers–patients.

2.2. Instruments

2.2.1. Disease severity

Disease severity was assessed using the ALSFRS – R, self-complete version. The ALSFRS – R comprises four subscales: bulbar function, fine motor function, gross motor function and respiratory function. Maximum score is 48; higher scores denote higher functioning. To facilitate clinical interpretation of findings, ALSFRS – R scores were categorized into four stages of severity: mild (37–48); moderate (25–36); severe (13–24); and very severe (0–12).

2.2.2. Activities of daily living

ADL were assessed via a modified version of the Disability Assessment of Dementia (DAD). The presentation of questions was made easier for the carer to tick their answers; the content was not altered. The DAD is a specific measure of ADL performance, comprising 40 questions: 17 pertaining to BADL, such as dressing, eating, and hygiene tasks and 23 related to IADL, such as meal preparation, managing finances and medication. One of the main advantages of the DAD is that each task is subdivided into initiation, planning and execution, which avoids floor performance on patients with marked motor impairment. Most well-known and regularly used ADL scales are based on physical impairment, overlooking frontal aspects of ADL performance such as initiation and planning. Moreover, DAD scores are corrected to pre-morbid functioning (that is, questions that are not applicable are not considered for the final score, which avoids gender bias). For instance, if patient is physically unable to cook at present, but had never cooked regularly prior to disease onset, this task is not scored. Maximum score is 100; lower scores represent more disability. The DAD was completed by an informant/carer.

2.2.3. Behavioural change

Changes in behaviour were measured by the Cambridge Behavioural Inventory Revised (CBI – R) scale score,[13,14] which comprises 10 domains (memory/orientation; everyday skills; self care; abnormal behaviour; mood; beliefs; eating habits; sleep; stereotypic and motor behaviour; motivation). For the purposes of this study we used four subscales, which reflected behavioural changes commonly associated with the frontal lobes: motivation, mood, stereotypical behaviour, and abnormal behaviour.[15] This selection was also based on their relevance to ADL performance.[6] The CBI – R rates frequency of behavioural change on a scale of 0 to 4, with higher scores indicating severe behavioural changes (0, no impairment; 1, a few times per month; 2, a few times per week; 3, a daily occurrence; 4, constant). CBI scores were corrected according to the number of questions per domain so that all four domains could be compared between them. In terms of severity, domain scores were further classified into mild (0–25); moderate (26–50); severe (51–75); and very severe (76–100)[15] to facilitate clinical interpretation of behavioural changes. The CBI – R was completed by the carer.

2.3. Statistical analysis

Data were analyzed using the Statistical Package for the Social Sciences version 17.0 (SPSS, Chicago, IL, USA). *A priori* variables were plotted and checked for normal distribution by Kolmogorov–Smirnov tests. One-way analyses of variance (ANOVA) were conducted to verify effects of ALSFRS – R stages on DAD total, BADL and IADL scores, with *post hoc* tests to compare the differences between stages. The Mann–Whitney *U*-tests were performed to compare bulbar and limb onset group differences on BADL, IADL as well as in each ADL activity, given the non-normal distribution of BADL scores.

3. Results

The baseline characteristics of our group are described in Table 1. The mean group age was 63.6 years (standard deviation = 9.1) and 63.6% of patients were male. Distribution of the patients by motor onset showed a predominance of limb ($n = 59$) onset over bulbar onset ($n = 23$). There was a higher proportion of male patients, and a tendency for older patients to have the bulbar subtype.

3.1. Disease severity

The ALSFRS – R scores were divided into four stages: mild (37–48), moderate (25–36), severe (13–24) and very severe (0–12). This is novel and was done to facilitate the interpretation of results. Of these stages, 33% of patients were in the mild stage; 48% in the moderate; 14% in the severe and 4% in the very severe stages.

3.2. Activities of daily living

As expected, patients with high ALSFRS – R scores had higher DAD scores, suggesting that functional disability increased with the severity of MND as measured by the ALSFRS – R scale (Fig. 1A). An ANOVA demonstrated a major effect of ALSFRS – R stage ($F = 11.883$, $p < 0.001$). *Post hoc* tests revealed that the severe and

Table 1
Comparison of patients with motor neuron disease subtypes (limb and bulbar) for demographics, disease severity and the Disability Assessment of Dementia (DAD) scale

	Group total ($n = 82$)	Limb ($n = 59$)	Bulbar ($n = 23$)	Limb *versus* bulbar
Male (%)	63.6	71.2	41.2	*
Age (years)	63.8 (9.1)	62.6 (9.3)	67.5 (7.4)	$p = 0.051$‡
Education (years)	12.8 (2.8)	13.2 (9.3)	11.9 (2.5)	n.s.‡
Disease severity (ALSFRS – R) ($n = 76$)	31.8 (8.9)	32.2 (9.1)	30.1 (8.3)	n.s.‡
DAD ($n = 82$)	72.5 (27.4)	71.7 (26.7)	81 (20.9)	n.s.*

ALSFRS – R = Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised score, n.s. = not significant.
* Mann–Whitney *U*-test, $p < 0.05$.
‡ Independent samples *t* test.