Disability profile and the factors affecting functional outcome in Malaysian Motor Neurone Disease population

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• Background & Objectives

Motor neurone disease (MND) is a progressive neurogenerative condition that affects primarily the motor neurons, leading to muscle weakness. The accurate knowledge on the individual spectrum of disabilities and the course of the disease is important for timely medical and rehabilitation management of patients with MND in terms of clinical prediction and clinical decision making. The objectives of this study are to identify the disability profile, functional outcome, fatigue and the correlation between fatigue and domains of functional decline in Malaysian MND population.

• Methods

Study design: A prospective cross-sectional study. **Population:** 30 MND patients undergoing rehabilitation programme at University Malaya Medical Centre (UMMC) were consecutively enrolled between January 2017 till April 2018. **Inclusion criteria:** 1) Age more than 18 years 2) Ability to participate in rehabilitation program which includes physiotherapy / occupational therapy and speech therapy sessions. **Exclusion criteria:** 1) Patients with severe cognitive impairment 2) Patients with other substantial medical, neurological or psychiatric disorder 3) Patients who are bed bound 4) Patients on assisted breathing . **Intervention:** Each patient completed a checklist to report the symptoms and problems affecting daily life. Amyotrophic Lateral Sclerosis Functional Rating Scale-revised (ALSFRS-R) and Fatigue Severity Scale (FSS). The ALSFRS-R was repeated again after 6 months to measure the functional decline. **Statistical analysis:** Descriptive statistics were used to represent the disability profile. Factors associated with functional status decline were analyzed using Mann Whitney U test, Kruskal Wallis test and independent t-test.

• Results

30 patients who fulfilled the inclusion and exclusion criteria participated in this study with 3 deaths during the course of the study. The mean age was 60 years with men more affected that women (3:1) *(Table 1)*. The main symptoms reported were weakness (93%), weight loss (83%) and fatigue (73%) *(Figure 1)*. Most had difficulties in performing domestic chores (77%) and engaging in social life (73%) *(Figure 2&3)*. No statistically significant factors were found to be associated with functional decline among the Malaysian MND patients *(Table 2)*. Decline in respiratory function was shown to be statistically significant among the fatigued MND patients (p=0.032) *(Table 3)*

Variable	Average/frequency
Age [mean ± SD (range)]	**59.73±11.74 (35-88)**
≤54	10 (33.3)
55-64	7 (23.3)
≥65	13 (43.3)
Gender (n; %)	
Male	22 (73.3)
Female	8 (26.7)
Ethnicity	
Malay	7 (23.3)
Chinese	19 (63.3)
Indian	4 (13.3)
Comorbidities (n; %)	
None	14 (46.7)
1	8 (26.7)
≥2	8 (26.7)
Site of Onset (n; %)	
Bulbar	6 (20.0)
Upper Limb	9 (30.0)
Lower Limb	15 (50.0)
Age of symptom onset [mean ± SD (range)]	**55.73±13.60 (21-82)**
≤54	12 (40.0)
55-64	11 (36.7)
≥65	7 (23.3)
Duration between symptom onset to diagnosis (in mouths) [mean ± SD (range)]	**16.10±19.57 (1-108)**
≤12 months	17 (56.7)
>12 months	13 (43.3)
Usage of Riluzole (n; %)	
Yes	10 (33.3)
No	20 (66.7)
Initial FSS score [(mean ± SD (range)]	**38.67±14.40 (9-63)**
FSS Score <40	13 (43.3)
FSS Score ≥40	17 (56.7)

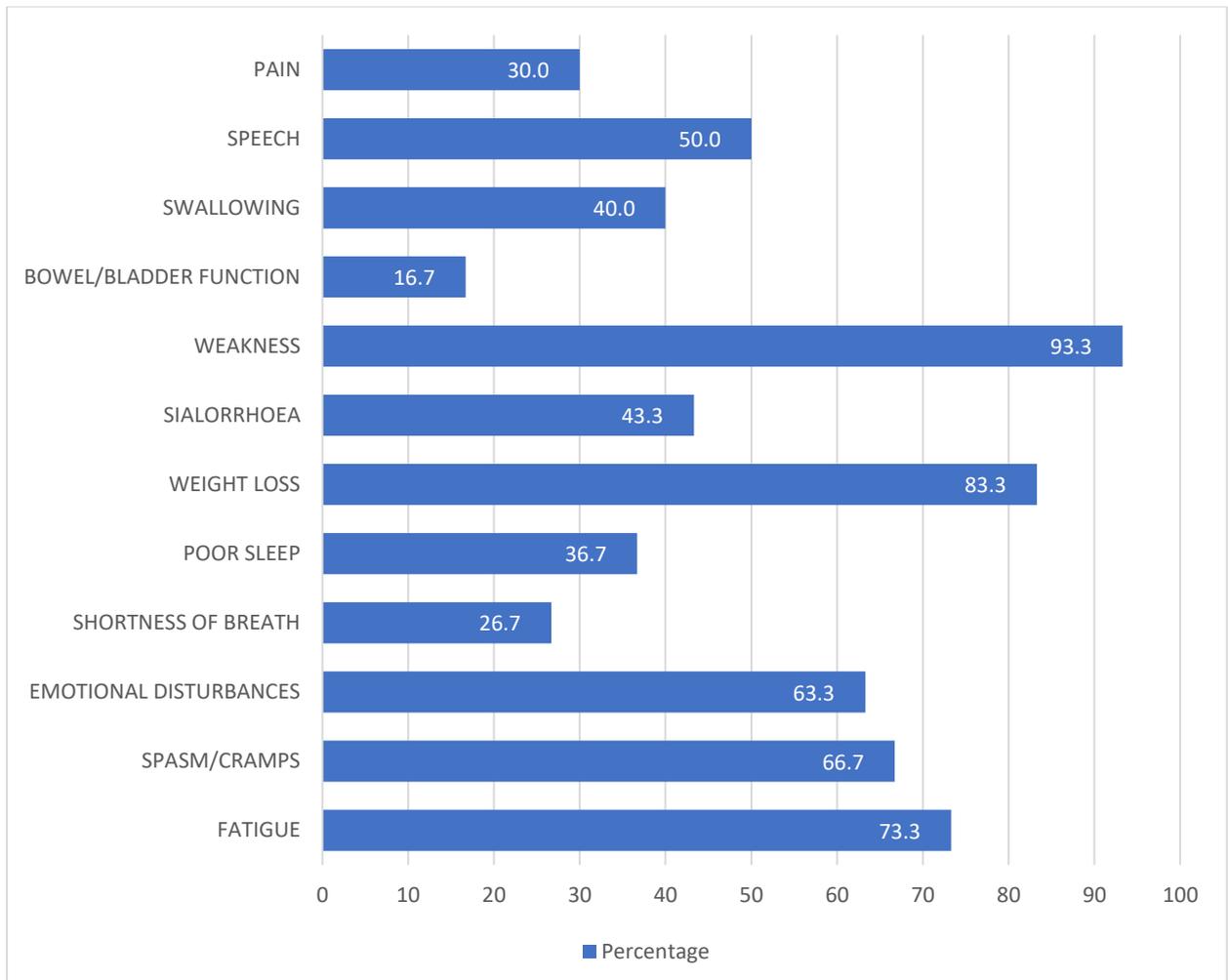*Table 1: Demographics and clinical characteristics of MND participants*

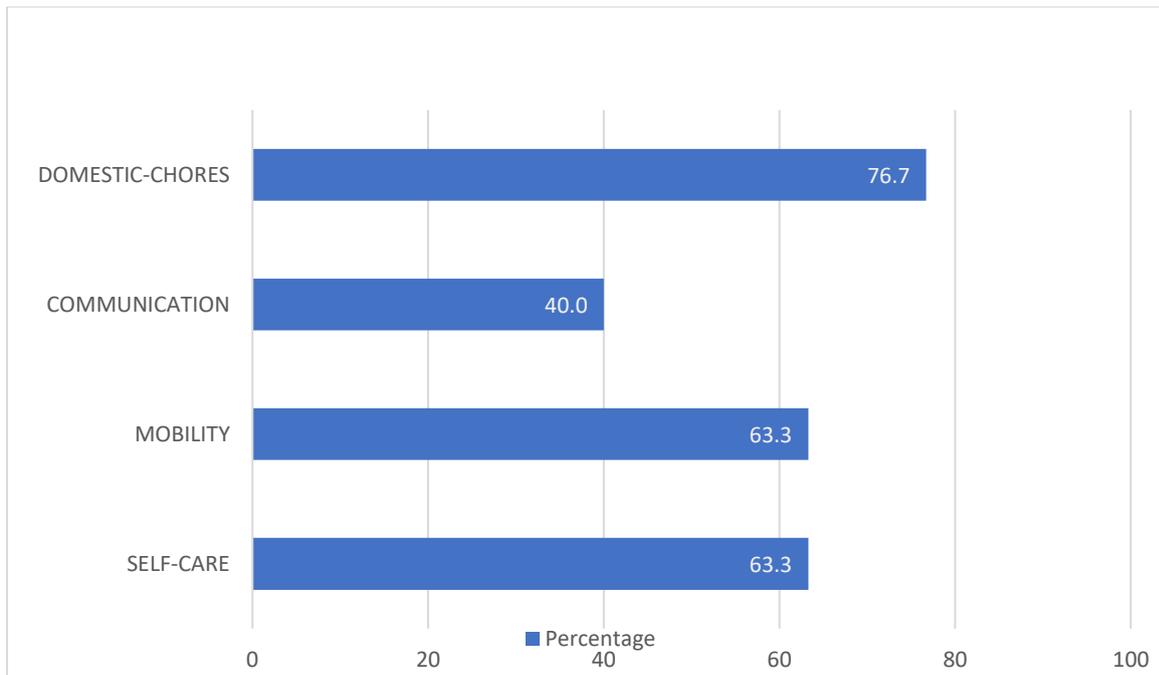*Figure 1: Impairments (body structure and function) of MND participants*

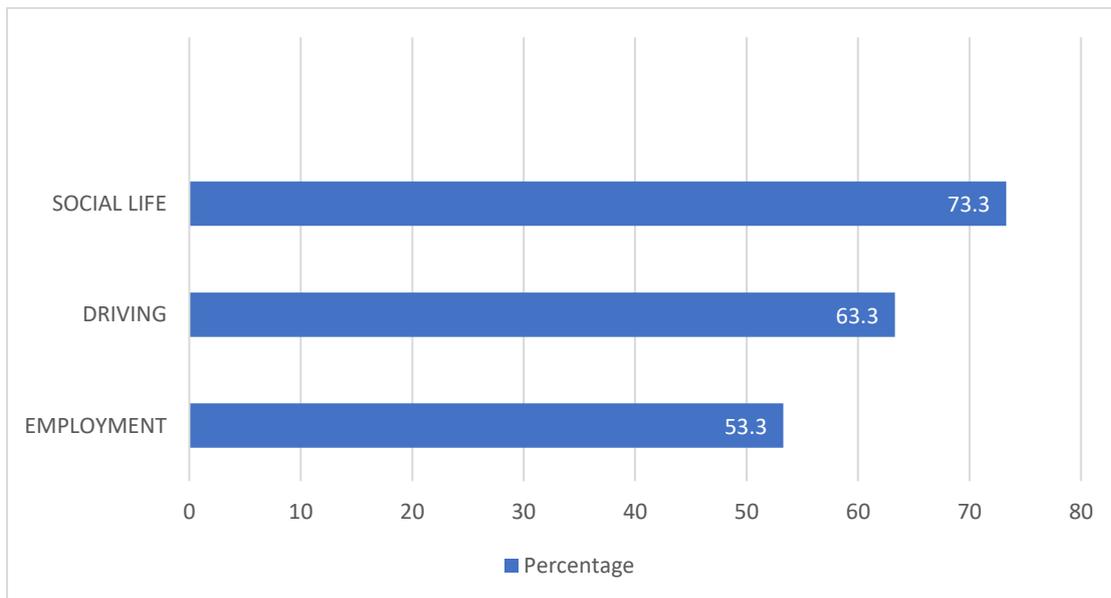*Figure 2: Activity limitations of MND participants*

Figure3: Participation restriction of MND participants

Variable	Decline in mean ALSFRS-R scare in 6 months [(mean decline = 6.85 (SD 4.5))			
	Mean	Standard Deviation	Median	*p* value
Gender				
Male	5.70	3.29	4.00	0.179
Female	10.14	6.15	12.00	
Ethnicity				
Malay	6.14	2.97	5.00	0.984
Chinese	6.83	4.73	4.50	
Indian	9.50	9.19	9.50	
Age				
54 and less	6.60	2.76	6.50	0.548
55-64	6.00	5.59	4.00	
65 and above	7.55	5.48	4.00	
Comorbidities				
None	6.85	4.04	6.00	0.722
1	6.71	4.23	4.00	
≥2	7.00	6.22	4.00	
Site of onset				
Bulbar	6.80	4.60	6.00	0.997
Upper limb	7.33	5.32	5.00	
Lower limb	6.54	4.29	4.00	
Age of symptom onset				
54 and less	6.25	2.63	5.50	0.447
55-64	6.44	5.81	4.00	
65 and above	8.67	5.72	7.50	
Duration between symptom onset to diagnosis				
≤12 months	6.75	4.80	4.00	0.716
>12 months	7.00	4.36	5.00	
Usage of Riluzole				
Yes	6.88	5.67	4.00	0.658
No	6.84	4.15	6.00	
Fatigue severity Scale (FSS)				
<40	4.75	2.22	4.00	0.053
≥40	8.53	5.25	6.00	

Table 2: Association of demographic factors and clinical characteristics to decline in ALSFRS-R score in 6 months

Functional Domains within ALSFRS-R	Initial Fatigue Severity Score	n	Decline in ALSFRS-R score in 6 months			
			Mean	Std. Deviation	Median	p value
Bulbar domain	Less than 40	12	-1.83	2.79	-1.00	0.867
	40 and above	15	-1.73	2.12	-1.00	
Fine Motor domain	Less than 40	12	-1.75	1.60	-1.50	0.063
	40 and above	15	-3.07	1.91	-3.00	
Gross Motor domain	Less than 40	12	-1.58	1.88	-1.00	0.277
	40 and above	15	-1.93	1.34	-2.00	
Respiratory domain	Less than 40	12	-0.33	1.16	0.00	0.032
	40 and above	15	-2.20	3.12	-2.00	

Table 3: Effect of fatigue on the different functional domains of the ALSFRS-R

• **Discussion**

Disability Profile

In our study, we found that weight loss was one of the main reported symptoms amongst the MND population. Factors that likely contribute to weight loss include loss of appetite, dysphagia, dyspnea, emotional disturbance and progressive muscular atrophy. Weight loss reflects nutritional status decline and it is considered to be a negative predictor of survival in MND patients. It is vital to perform routine nutritional status assessment for all patients and rendering nutritional therapy if necessary. In contrast to previous studies, our patients had more issues in performing domestic chores as compared to mobility and self-care. The availability of many ambulatory and adaptive aids and equipment in recent years has assisted patients in aspects of mobility and personal care. Performance of domestic chores can be enhanced by adapting one's environment or adding major installations. Social life among MND patients was largely affected by social embarrassment limited mobility, speech and communication difficulties, Social isolation can lead to emotional disturbances and depression. The latter features require further attention to allow patients to have a more meaningful quality of life.

Factors affecting the course of functional status

We failed to identify significant demographic and clinical characteristics that are associated with functional decline in our MND patients. The progression rate and the course of disease in MND was heterogenous.

Fatigue and its effect on functional outcome

Fatigue in MND is attributed to physical causes due to motor neuron degeneration. In addition, features such as depression, sleep problems, respiratory insufficiency and weight loss may contribute to fatigue. There was no statistically significant relationship between fatigue and functional deterioration in MND population. However, further analysis of the different functional domain within the ALSFRS-R demonstrated that fatigue was associated with worsening respiratory domain. Fatigue is prevalent in MND and has a significant impact

on quality of life, thus it is important to overcome this impairment. In terms of rehabilitation, patients should be advised to pace their daily activities and exercise regime to prevent fatigue from setting in. Focus should be shifted on providing routine pulmonary rehabilitation to MND patients especially those with fatigue to dampen the progressive deterioration of respiratory function.

Study Limitations and Future Recommendation

This was a single centered study with a small sample size limited by the time, the rarity and high mortality rates that comes with MND, The functional assessment was only done twice within 6 months which is too short a time to gauge one's functional course, In future, further studies recruiting larger sample size with frequent functional assessment throughout a course of prolonged duration are required to establish the factors affecting the status of functional decline among MND patients to facilitate rehabilitation management. Future studies should investigate the impact of rehabilitation programs on disease progression.

• Conclusion

Framework of International Classification of Functioning, Disability, and Health should be explored in MND populations to provide the best multidisciplinary care through the 'neuropalliative rehabilitation' model, In terms of rehabilitation, a paradigm shift is needed to provide pulmonary rehabilitation to MND patients especially the ones with fatigue to dampen the progressive deterioration of respiratory function enhancing the quality of life.

• References

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