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ORIGINAL ARTICLE

Physical Activity in People With Motor Neuron Disease: Validity of the Physical Activity Scale for the Elderly as a Measuring Tool

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Abstract

Objective: This study aimed to investigate whether the physical activity scale for the elderly (PASE) is a valid tool in measuring physical activity (PA) in people with motor neuron disease (MND) and to identify the demographic and clinical factors that predict PA participation in this population.

Design: A prospective, observational study involving 100 ambulant participants with MND.

Setting: This study was conducted at a multidisciplinary specialist MND clinic. The clinic is fully funded by the local public health system and patients receiving care here are not expected to pay for their consultation.

Participants: 190 patients with MND who had a physiotherapy appointment at the specialist clinic between July and October 2018 were screened. Of these, 100 participants (mean age 67 years [SD=12], 64% [n=64] men) who were ambulant (with or without assistance) were recruited (N=100).

Interventions: Not applicable.

Main Outcome Measures: PASE questionnaire, amyotrophic lateral sclerosis functional rating scale—Revised (ALSFRS-*r*), forced vital capacity (FVC).

Results: The results showed that engagement in PA is generally low, with median PASE score of 57. The PASE had fair-moderate correlation with ALSFRS-R total scores (rho=0.607; $P<.000$) and FVC (rho=0.250; $P=.030$). Standard multiple regression analyses showed that disease severity (ALSFRS-R total score) was the strongest predictor of PA levels ($\beta= 0.54$; 95% confidence interval 0.02,0.06). The most frequently selected physical activities of choice for people with MND were activities around their homes and the biggest barrier to participation is fatigue.

Conclusion: Present findings suggest that the PASE can be used to measure PA participation in people with MND. Details about activity of choice and barriers to participation present important considerations in designing exercise programs in this population to maximize compliance and therefore effectiveness.

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Motor neuron disease (MND) is a rare and fatal neurodegenerative condition characterized by progressive weakness, dysarthria, dysphagia, and respiratory compromise.[1] Clinically this manifests as progressive impairment in activities of daily living and mobility, communication deficits, swallowing and breathing difficulties, and fatigue.[2]

Current management of patients with MND focuses largely on symptom management and maximizing quality of life. Evidence suggests that adults with physical disabilities have low levels of leisure time physical activities (PAs) participation.[3] The reduced activity that occurs as a result of chronic disease or disability can lead to a cycle of secondary deconditioning, with impairments causing physical deterioration and further subsequent reductions in activity.[4]

While the consequences of inactivity are well documented in neurologic conditions such as stroke and Parkinson's disease (PD), less is known about exercise behavior of people with MND. Several systematic reviews have demonstrated that exercise is a safe option for people with MND; however, the type, intensity,

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and frequency or when to initiate exercise therapy remains largely unknown.[5-7] Mild to moderate endurance training was reported to improve survival in pre-clinical mouse models[8]; however, currently there are no clinical evidence supporting this in ALS patients. Research into this topic may be difficult because of the heterogeneity and rapidly progressing nature of the condition.

Despite the lack of evidence, exercise is recognized as an important modality in managing some of the symptoms associated with MND. The recent MND National Institute for Health and Care Excellence (NICE) guideline recommends that an exercise program should be considered to optimize function and improve quality of life, and to *"choose a programme that is appropriate to the person's level of function and tailored to their needs, abilities and preferences"*.[9] However, the needs, abilities, and preferences in this population is largely unknown. In our clinical experience, nonspecific encouragement to stay active, to engage in physical activities but to avoid over exertion is typically provided by health professionals.

The PASE (physical activity scale for the elderly), designed specifically for older adults, has been used in numerous studies exploring PA in people with neurologic conditions such as PD[10] and stroke.[11] However, no published information describing PA, as measured using the PASE in people with MND has been reported.

This study aimed to assess the use of the PASE as a tool to measure PA participation in ambulant people with MND by examining its association with measures of disease severity and lung function. The demographic and clinical factors that predict PA levels were examined, as well as the types of activities people with MND choose to participate in.

Methods

Study design

A prospective, observational study (Registered: ACTRN126 18000889257) was undertaken at a state-wide multidisciplinary specialist MND clinic. The clinic treats adults with MND throughout the state from both metropolitan and rural settings. Ethical approval was obtained from Calvary Health Care Bethlehem Research Ethics & Ethics Committee (Number 18041201).

Participants

All patients with MND who had a physiotherapy appointment at the specialist MND clinic between July and October 2018 were screened. Of these, participants who were ambulant (with or without assistance) were invited to participate in the study and provided written informed consent if interested. Participants with comorbidities affecting PA participation were excluded from the study.

List of abbreviations:

ALSFRS -R	**amyotrophic lateral sclerosis functional rating scale —revised**
FVC	**forced vital capacity**
MND	**motor neuron disease**
NICE	**National Institute for Health and Care Excellence**
PA	**physical activity**
PASE	**physical activity scale for the elderly**
PD	**Parkinson's disease**

Outcome measures

PASE

The PASE is a brief and easily administered questionnaire into PA participation in the last 7 days. It includes questions regarding engagement with a range of physical activities of different intensities and hours committed to these activities. Type of activity and activity frequency are weighted to calculate a total PASE score, with higher scores indicating greater PA participation. The PASE demonstrates good convergent validity in comparison with Actigraph Monitors, physiological and performance characteristics of exercise for example, peak oxygen uptake[12] and accelerometer data.[13] It has also been shown to have good inter-rater reliability.[12,13] Information about the types of activities people with MND choose to do were extracted from the PASE to observe for common themes.

Amyotrophic lateral sclerosis functional functional rating scale—revised

The amyotrophic lateral sclerosis functional functional rating scale—revised (ALSFRS-R) is a self-reported questionnaire of current function. It comprises 12 items categorized into 4 equal domains: fine motor, gross motor, bulbar, and respiratory function. Each item has 5 possible answers (0=unable to do and 4=normal function). The scores are tallied into sub-scores for each domain (0-12), which can then be combined to give a total score ranging between 0 and 48. The ALSFRS-R is the most widely used marker of disease progression in MND currently as it showed a strong correlation with disease progression and survival.[14] Franchignoni et al suggest that the ALSFRS-R should be considered as a profile of sub scores rather than a single total score.[14]

Forced vital capacity

Forced vital capacity (FVC) is a measure of inspiratory muscle strength and is expressed as percentage of expected value. FVC has been shown to be a predictor of disease progression and survival.[15,16]

Data collection

All data except for FVC were collected by the treating physiotherapist. Demographic data (age, sex, exercise behavior pre-diagnosis, time since diagnosis, and MND phenotype) were collected the first time the participant was seen during the data collection period. Falls history was reported as the number of falls in the 6 months prior to the data collection date.

ALSFRS-R and PASE were collected through an interview either face to face or over the phone. Participants were asked additional multiple choice questions about what motivates them to exercise and factors which prevents them from participating in PA or exercise.

Seated FVC was taken from the participant's routine lung function test if it was completed or would be conducted within 4 weeks of the data collection date. Where FVC was not available, it was measured on the day of data collection by the physiotherapist using the Micro Vitalograph[a].

Data analysis

Descriptive statistics were used to present baseline characteristics. The Shapiro-Wilk test and visual inspection of histograms were used to evaluate data distribution to identify parametric vs non parametric data.

Correlations between key outcomes (ALSFRS-R total score, ALSFRS-R sub-scores, FVC, PASE) were determined using Pearson r or Spearman rho depending on the distribution of the data. Determination of the strength of any relation was performed using the following guidelines from Cohen[17]: small, r=0.10 to 0.29; medium, r=0.30 to 0.49; large, r=0.50 to 1.0.

Standard multiple linear regression analysis was performed to examine whether demographic characteristics (eg, age and sex), disease severity (ALSFRS-R scores and gait status) or lung function (FVC) contributed to PA levels. The dependent outcome variable was PA levels as measured by the PASE total score, and the independent variables were age, sex, disease duration (months since diagnosis), disease severity (ALSFRS-R scores), lung function (FVC), and gait status (independent or dependent). All independent variables were treated as continuous variables except for sex and gait status that were coded into categorical variables. In order to ensure that the assumptions for multiple regression analyses were met, preliminary screening using residual plots was used. An initial evaluation led to a log transformation of the PASE total score in order to reduce skewness as well as improve the normality of the residuals. SPSS v24 (SPSS Inc, Chicago, IL) was used for all statistical calculations. The level of significance was set at P<.05.

Types of PA were categorized, examples provided, and percentage calculated from free text responses to PASE surveys. After interviews, if participants acknowledged that there were barriers to PA participation, fixed choice options were presented to best explain the barrier (fatigue, lack of time, advised not to, does not know what to do, other) and enabler (personal enjoyment, reduce disease progression, advised to by health professionals, other). More than 1 response could be chosen. Response frequency was calculated.

Results

One hundred ninety participants were screened and 100 ambulant participants were recruited into the study, ranging in age from 29 to 86 years (mean 67 years, SD=12) with 64 men (64%). Bulbar was the most prevalent clinical phenotype (34%) and primary lateral sclerosis was the least represented phenotype (7%) in the cohort (table 1).

Correlation analysis

Because of the underlying skewed distribution, Spearman's rank correlation coefficient was used to assess the association between the PASE, ALSFRS-R, and FVC. There were significant large positive correlations between PASE and ALSFRS-R total scores (rho=0.596; P<.001), fine motor sub-score (rho=0.627, P<.001) and gross motor sub-score (rho=0.532, P<.001). There was a medium positive correlation between PASE and breathing subscore; however, there was no significant correlation between bulbar sub score and PASE (table 2).

Predictors of PA

For PA levels as measured by the PASE score, multiple regression analysis showed that disease severity (ALSFRS-R total score) was the strongest predictor β =0.54; 95% confidence interval 0.02-0.06; P<.001) (table 3). This model accounted for 25% of the variance in PA levels, with disease severity (ALSFRS-R) explaining 18% of the variance in PASE score.

Lung function (FVC) was not found to be a significant predictor of the PASE score, contributing less than 1% to the regression model. The remaining variables also only contributed 1% or less to the model.

Types of physical activities

Table 4 indicates the number of participants engaging in different types of activities. Walking outside the home was the most frequent response followed by house work. Participation in exercise groups and sports were the least frequent responses.

Barriers to PA participation

Those who identified that there was a barrier to PA participation (n=94) were presented with 4 potential barriers to select from (table 5). Results suggest that fatigue was the biggest barrier for people with MND to participate in physical activities.

Motivators to exercise

Participants were presented with potential motivators to select from (table 6). Results indicate that most people exercise because they enjoyed the physical activities.

Discussion

Study findings suggests the PASE can be used as a tool to measure self-reported PA levels in ambulant adults with MND. The PASE does not require any specialized equipment, can be completed in person, over the phone or written mail out, and takes less than 10 minutes making it a quick and easy tool to use in both the clinical and research setting. Potential use for the PASE may be to track PA participation over time or to measure the effect of an intervention aiming at improving PA participation. The PASE provides therapists with a detailed account of the patient's PA pattern, which would be useful for therapists to tailor an exercise program to meet the patient's interest and therefore maximize compliance.

Looking at PA participation, findings suggest that it is very low in people with MND-median PASE score of 51.79 (interquartile range: 0-318.11), compared with 175 in healthy controls,[10] 162.6 in people with early PD,[10] and 99 in people who have had a mild stroke.[11] Regression analysis showed that PA participation is most strongly associated with MND disease severity (ALSFRS-R total score). Similar findings were found by Tsukita et al in people with PD.[18]

Closer analysis of ALSFRS-R sub-scores, the biggest correlation was with the gross and fine motor domains. MND is characterized by progressive motor weakness which spreads throughout the body. This leads to gross motor function decline including gait changes such as foot drop and decreased stride length, poor balance, and postural instability.[19] In the early days, this may mean reduced engagement in physically demanding activities such as

Table 1 Participants characteristics

N=100	Mean (SD, Range)
Age (y)	67 (12, 29-86)
Sex (M) (n)	64
Months since diagnosis	32 (23, 5-113)
Participated in regular sport or exercise pre diagnosis (n)	76
ALS subtype (n)	
Bulbar	34
ALS (cervical onset)	29
ALS (lumbar onset)	21
Flail arm	8
PLS	7
Missing	1
ALSFRS Median (IQR)	
Total score	33.50 (13-48)
Bulbar	10 (1-12)
Fine motor	8 (0-12)
Gross motor	6 (2-12)
Breathing	10.5 (2-12)
Falls reported in the previous 6 mo (n)	
0	60
1-2	26
3-5	9
>6	5
Independent gait (no aids, no assistance) (n)	44
FVC (% age matched normal)	80.7 (23.8, 16-121)
PASE total score Median (IQR)	51.79 (0-318.11)

Abbreviation: IQR, interquartile range; PLS, primary lateral sclerosis.

Table 2 Correlations between disease severity, lung function and physical activity

	ALSFRS-R Total Score	FVC	PASE Total Score
FVC (percentage of age-matched normal)	0.536*	-	0.232†
PASE total score	0.596*	0.232†	-
ALSFRS-R total score	-	0.536*	0.596*
Bulbar sub-score			0.049
Fine motor sub-score			0.627*
Gross motor sub-score			0.532*
Breathing sub-score			0.365*

NOTE. Spearman's rho nonparametric correlations.
* Significant at $P<.01$.
† Significant at $P<.05$.

sports, running, or cycling. As more body areas are affected and weakness becomes more pronounced, some are unable to continue working, become more reliant on carers for support with household chores, and eventually require assistance with personal care tasks. In other words, as their MND progresses, gross and fine motor function decreases and their ability to perform physical tasks diminishes resulting in a low PASE score.

At a certain point, people with MND are unable to do exercise or perform PA tasks safely on their own, and therefore need assistance from a carer. By this stage, they normally also rely on carers for assistance with other aspects of their life. This includes domestic and personal care tasks, maintaining good nutritional intake and managing daily routines including medical appointments. These tasks usually fall on the shoulders of families and friends who are concurrently dealing with other life responsibilities. Galvin et al 2016 reported that on average, caregivers spend 47 hours per week providing care, with 44% of caregivers working while providing care.[20] The burden of care increases in parallel with disease severity.[21] Managing competing priorities and dealing with declining physical function were identified as significant contributing factors to increasing burden of care.[20] When competing with other necessary daily tasks, assisting with participation in physical activities becomes less of a priority for carers. Clinically, it is observed that by this stage, most of the physical activities performed are necessary such as getting out of bed, walking to the toilet, and maybe stretches for joint pain relief and comfort. Additional PA is not achievable. In many cases, there is simply no time left in the day for them to be supported to exercise in a safe way.

The finding that the PASE score correlated more strongly with the fine motor than gross motor activities was unexpected. However, all the physical activities, except for walking, captured by the PASE involves the upper limbs, such as gardening, housework, and sporting activities. This may explain the strong correlation between PASE score and fine motor sub-score. This finding may have significant clinical implications as people with MND may be forced to limit the types of physical activities they engage in because of difficulties manipulating items (whether that be in the garden, the house, or the gym), not because they lack the overall muscle strength to complete the task.

Interestingly, gait status or self-reported falls were not predictors of PA participation as measured by total PASE score. One explanation for this may be the participants' reluctance to accept gait aids recommendations. This means that they may be self-limiting their walking/PA participation for a period, before accepting the use of a gait aid. Once they have made the mental adjustment to accept their disease progression and accept the use of a gait aid, PA participation may in fact increase. Similarly, people more at risk of falls may limit themselves, as a strategy to reduce falls risks, and therefore report no falls. These relations are complex and further study into this topic would provide insightful information.

Fatigue is 1 of the most commonly reported symptoms of MND[22] and worsens as disease progresses.[22] Clinically speaking, fatigue is often reported as a reason for reducing PA participation. In fact, 62% of study participants reported fatigue to be the main barrier limiting PA participation. Despite being common, fatigue is poorly understood. The feeling of fatigue was found to be determined by dyspnea-related distress, rather than any objective measure of respiratory function.[23] It was also noted that participants who are treated with noninvasive ventilation experience fatigue more intensely than those who are not.[23] Although fatigue and dyspnea were not specifically explored, our results showed a significant correlation between the PASE score and breathing subscore which contains items about dyspnea and noninvasive ventilation usage. This suggests that there may be an inverse relation between fatigue and PA participation.

Our results showed that in the MND population, there was a small correlation between FVC and PA. When other covariates (eg, age, disease severity) were included in the multiple regression model, lung function was not a significant predictor of PA levels. This was surprising, as we expected lung function as measured by FVC to play more of a role in determining the amount of PA

Table 3 Standard multiple regression of factors associated with physical activity levels

Model	B	β	R^2	*t*-Value	95% CI	*P* Value
Constant	0.76					
ALSFRS-R total		0.54	0.177	4.37	0.02, 0.06	0.000*
FVC (percentage of age-matched normal)		-0.11	0.008	-0.93	-0.01, 0.00	0.354
Age		-0.08	0.006	-0.79	-0.01, 0.01	0.432
Sex		0.11	0.011	1.09	-0.08, 0.28	0.279
Disease duration		-0.08	0.005	-0.76	-0.01, 0.00	0.448
Gait status		-0.09	0.007	-0.90	-0.26, -0.10	0.374
R^2=0.30 Adjusted R^2=0.25						

Abbreviations: β, standardized regression coefficients; CI, confidence interval; R^2, unique contribution of each predictor variable to the total variance in overall PASE score.
* $P<.05$.

Table 4 The types of physical activity engaged in by participants

Type of Physical Activity	Examples of Activity	Participation (%)
Walking outside the house	Walking to the shops, walking the dog, walking around the block	71%
Housework	Vacuuming, sweeping, packing and unpacking the dishwasher, hanging out clothes	51%
Home exercise: strengthening program	Exercise bike, upper and lower limb exercises, weights, theraband	24%
Home exercise: stretching program	Active and passive shoulder stretches, calf stretches	14%
Participation in sports	Bowls, golf, cycling	12%
Attends an exercise group	Yoga, Pilates, gym, hydrotherapy	10%

people engaged in. Perhaps this indicates that if someone has significant limb weakness, PA will be limited regardless of lung function. Disease severity and fatigue may play a bigger role in predicting PA participation than lung function for the reasons discussed above.

Looking at the types of activities that people with MND engage in, it was demonstrated that the most frequently selected activity was walking outside the home (71%), while the least frequent activity was attending an exercise group (10%). This combined with the barriers to PA participation as discussed above suggests that home-based activities are more easily achievable than center-based programs. This is an important factor when prescribing exercise in this population. The effects of the upper limb, and in particular hand weakness, may lead to limited participation is community activities. For example, upper limb weakness may preclude participation because of an inability to drive to a local gym or difficulties with dressing/undressing for hydrotherapy. Further research is needed to understand motivations and barriers to participation in exercise, both at home and in the community.

Limitations and future research directions

This was a single center study which may affect the generalizability of the results to other cohorts. However, participants were attending a state-wide clinic servicing people with MND living in a wide area with an approximate population of 6.5 million. Further, baseline demographic data are comparable with Talman et al's study which recruited 1834 participants from 10 specialist centers nationwide,[24] increasing the confidence in study results.

The PASE does not capture household ambulation, which may represent a floor effect regarding the measurement of PA in people with MND.

This study did not compare the total PASE score with PA data from an Actigraph. This would be the logical next step in validating the use of the PASE in this population.

Given that fatigue was recorded as the main barrier to PA participation by two-thirds of our cohort, it would be interesting to investigate how fatigue affects participation and how it can be better managed to support PA participation.

Table 5 Self-reported barriers to physical activity participation

Barriers*	**n**
Fatigue	61
Has no time/support to participate safely	14
Advised not to exert themselves	10
Doesn't know what they should/shouldn't do	9
No barrier identified	6
Bad weather	1

* More than 1 option could be selected. Five participants did not provide an answer.

Table 6 Self-reported motivators to physical activity participation

Motivators*	**n**
Enjoys exercising	47
Believes it will slow down disease progression	21
Advised by health professional to exercise	9

* More than 1 option could be selected. 25 participants did not provide an answer.

Conclusion

Present findings suggest that the PASE is a useful tool to measure PA participation in people with MND. It is a quick and easy tool that can be used in both clinical and research settings. As the PASE does not include walking within the home, combining the PASE with a household ambulation measuring tool may increase its sensitivity to the changes in physical activities in this population.

The most frequently selected PA for people with MND appears to be home-based activities and the biggest barrier to participation is fatigue These present important considerations in designing exercise intervention programs for this population to maximize compliance and therefore effectiveness.

Supplier

a. Micro Vitalograph; Model 6300, Vitalograph Ireland Ltd.

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Keywords

ALS; Amyotrophic lateral sclerosis; Exercise; MND; Motor neuron disease; Physical activity; Rehabilitation

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