Relationships between motor impairments and activity limitations in patients with neuromuscular disorders

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ABSTRACT

Aim: The strength and nature of the relationships between motor impairments and activity limitations assessed by the ACTIVLIM questionnaire were investigated in 245 patients with neuromuscular disorders.

Methods: Measures of motor impairments consisted of: (1) a grip strength test using a Jamar dynamometer, (2) a Manual Muscle Testing bilaterally performed in 18 muscle groups and (3) a gait speed spontaneously adopted by the patients using the 10 m timed walking test.

Results: Activity limitations were poorly correlated with grip strength in both hands ($r = 0.3$ and 0.36) and moderately correlated with gait speed ($r = 0.53$). Spearman's coefficients of correlation between the manual muscle testing and activity limitations were moderate to very poor ($\rho = 0.5$ to 0.17).

Conclusion: The relationships between motor impairments and activity limitations are not straightforward in patients with neuromuscular disorders, indicating that the activity limitations should be separately assessed and cannot be simply inferred from motor impairment measures.

The International Classification of Functioning, Disability and Health (ICF) proposed by the World Health Organization gives a framework to describe an individual's functioning, taking into account his health condition in three separate components[1]: (1) body functions and anatomical structures, (2) activity, defined as the achievement of daily activities and (3) participation, defined as involvement of the subject in a life situation. Problems or difficulties that a subject may have in each component are impairments, activity limitations and participation restrictions, respectively. Although these components are separately defined, they are related but not necessarily in a straightforward relationship.[2 3] Indeed, two individuals with the same level of impairments will not necessarily have the same level of activity or participation. The nature and strength of the relationships between impairments and activity limitations have been studied in patients with polyneuropathies[2] and in those with Duchenne muscular dystrophy[4 5] among patients with neuromuscular disorders (NMD).

The principal impairment in patients with NMD is deterioration of motor function, characterised by a progressive decrease in muscle strength.[6] The location and severity of motor impairments in patients with NMD vary widely according to the aetiology of the disease, as the origin of the muscle weakness, its physiopathology and the related symptoms depend on the type of NMD.[7] The activity limitations in patients with NMD were recently studied by developing and validating a new scale of activity limitations, the ACTIVLIM questionnaire.[8]

The purpose of this study was to assess the relationships between motor impairments and activity limitations in patients with NMD to verify whether the clinical tests frequently used to assess their motor impairments could be used to infer their ability in performing daily activities. In the same way, conventional treatments tend to maintain or improve joint mobility, muscle strength and endurance with the aim of providing patients the means of increasing their activity level.[1 9] The relationships between motor impairments and activity limitations were investigated in different diagnostic groups of NMD and in a wider sample of NMD without diagnosis distinction. This approach may therefore interest clinicians who take care of patients with a specific NMD but also those who follow large groups of patients in neuromuscular clinics.

PATIENTS AND METHODS

Patients

This multicentre study was approved by the medical ethics committees of the Université catholique de Louvain and the Katholieke Universiteit Leuven. Patients were recruited through the Neuromuscular Reference Centres of two university hospitals. Adult patients and parents of affected children gave written informed consent before the evaluation. A total of 245 patients (46 children from 6 to 16 years and 199 adults from 16 to 80 years) with a diagnosed neuromuscular disorder were assessed by two experienced examiners ($n = 127$ for examiner 1 and $n = 118$ for examiner 2). Six main diagnostic groups were identified from the sample, each including more than 5% of the sample: (1) Duchenne, Becker and limb girdle muscular dystrophy (DMD/BMD/LGMD) ($n = 45$, 6–72 years), (2) hereditary neuropathy (HN) ($n = 44$, 8–80 years), (3) myotonic dystrophy (MD) ($n = 37$, 16–72 years), (4) facio-scapulo-humeral dystrophy (FSHD) ($n = 12$, 12–67 years), (5) spinal muscular atrophy (SMA) ($n = 14$, 9–61 years) and (6) amyotrophic lateral sclerosis (ALS) ($n = 18$, 46–80 years). Patients not belonging to one of these groups made up the group "others" ($n = 76$, 6–80 years), including the rest of neuromuscular disorders such as post-polio syndrome, congenital muscular dystrophy, inflammatory or systemic neuropathies, or

Table 1 Patient sample (n = 245)

Characteristic	
Age (years) (mean (range))	42 (6–80)
Gender (n (%))	
Men	142 (58)
Women	103 (42)
Diagnosis (n (%))	
DMD/BMD or LGMD	45 (18)
HN	44 (18)
MD	37 (15)
ALS	18 (7)
SMA	14 (6)
FSHD	11 (5)
Other (eg, CM, CMD, PPS, ...)	76 (31)
Mobility level (n (%))	
Walking	169 (69)
Wheelchair bound	76 (31)

ALS, amyotrophic lateral sclerosis; BMD, Becker muscular dystrophy; CM, congenital myopathy; CMD, congenital muscular dystrophy; DMD, Duchenne muscular dystrophy; FSHD, facio-scapulo-humeral dystrophy; HN, hereditary neuropathy; LGMD, limb girdle muscular dystrophy; MD, myotonic dystrophy; PPS, post-polio syndrome; SMA, spinal muscular atrophy.

metabolic myopathy, for example. A description of the sample is given in table 1.

Motor impairment assessment

Patients were assessed individually in a quiet room during their multidisciplinary consultation at the neuromuscular centres. The tests were clearly explained to the patients and included strength measures and a measure of spontaneously adopted gait speed.

Strength measures comprised grip strength and manual muscle tests. The measure of grip strength has been reported as providing information about activity limitations[10 11] and even predicting functional limitations in the elderly.[12 13] Grip strength was measured with a Jamar dynamometer (Therapeutic Equipment Corporation, Clifton, New Jersey, USA) according to the procedure described by Mathiowetz and colleagues.[14] The average of the maximal force exerted on the dynamometer across three trials gave the measure of grip strength expressed in Newtons. There was 1 min of rest between each trial to avoid bias caused by fatigue. Grip strength was recorded for both hands.

Manual Muscle Testing (MMT) is a non-instrumented method for measuring muscle strength in patients with NMD.[15] Nine muscle groups of the upper limb (shoulder abductors and flexors, elbow flexors and extensors, wrist flexor and extensors, digit flexors, extensors and interossei) and nine muscle groups of the lower limb (hip flexors, extensors and abductors, knee flexors and extensors, ankle dorsiflexors and plantar flexors, toe flexors and extensors) were bilaterally tested using the six grade Medical Research Council Scale (0 = no movement; 1 = flicker of movement; 2 = movement of the joint when the effect of gravity is eliminated; 3 = movement through full range of the joint, against gravity; 4 = movement of the joint, against gravity and against added resistance; 5 = full strength).[16] Conditions of the MMT (positions of the patient and the examiner, application of manual resistance, stabilisation of the patient, etc) were standardised according to the procedure described by Kendall *et al*.[17]

The gait speed spontaneously adopted by the patient was reported to be a reliable index of locomotor impairment in patients with various pathologies of the lower limbs.[18] The spontaneous gait speed in our patients with NMD was obtained using the 10 m timed walking test. The 10 m test was considered as a reliable and valid measure in patients with amyotrophic lateral sclerosis,[19] immune mediated polyneuropathies[2] and in patients with Charot–Marie–Tooth neuropathy.[20] The patient was asked to walk a distance of 10 m at his own preferred and comfortable speed, starting from a standing position. The time taken to complete the task was recorded and the gait speed was expressed in m/s. Any help necessary to walk the 10 m was allowed. Fourteen per cent of patients used one or two crutches, 15% used an ankle–foot orthesis or orthopaedic shoes and 2% needed personal support to keep balance. Nevertheless, no data were recorded for wheelchair bound patients.

Table 2 The 22 items of the ACTIVLIM questionnaire ordered by decreasing difficulty

	Item
A	Hopping on one foot
B	Carrying a heavy load
C	Running
D	Walking more than 1 km
E	Walking upstairs
F	Standing for a long time
G	Stepping out of a bath tub
H	Walking downstairs
I	Taking a bath
J	Putting on a backpack
K	Dressing one's lower body
L	Walking outdoors on level ground
M	Getting into a car
N	Taking a shower
O	Wiping one's upper body
P	Putting on a T-shirt
Q	Hanging a jacket on a hat stand
R	Sitting on the toilet
S	Washing one's upper body
T	Opening a door
U	Closing a door
V	Washing one's face

Activity limitations assessment

Activity limitations were assessed with the ACTIVLIM questionnaire.[8] This questionnaire assesses the difficulties a patient may have in executing daily activities.[1] It contains 22 daily activities designed for both children and adults with NMD (table 2). The children's parents and the adult patients were asked to provide the difficulty they perceived in performing each activity on a three level ordinal scale (0 = impossible, 1 = difficult or 2 = easy). Participants were instructed that the activities should be completed without human help. The ordinal total score obtained on the ACTIVLIM questionnaire was subsequently transformed into an interval level measure of activity limitations according to the Rasch model.[21] The activity limitations scale has a constant measurement unit, called logit; the higher the value in logits, the higher the patient's activity level. As the measures are linear, they can be treated as continuous variables.

Statistical analysis

The scores of grip strength and gait speed were transformed into standardised z scores according to normative data available in

Table 3 Prevalence of motor impairments according to type of NMD

	DMD/BMD LGMD (%) (n = 45)	HN (%) (n = 44)	MD (%) (n = 37)	FSHD (%) (n = 11)	SMA (%) (n = 14)	ALS (%) (n = 18)	Other (%) (n = 76)	Total sample (%) (n = 245)
Grip strength (right)	73	48	87	55	77	67	46	60
Grip strength (left)	78	40	81	18	77	61	35	53
Gait speed in walking patients	69	63	52	75	50	45	65	60
Upper limb proximal muscles								
Shoulder flexion	78	8	15	100	75	53	46	44
Shoulder abduction	76	11	17	100	92	56	43	45
Elbow flexion	72	8	3	36	75	29	27	31
Elbow extension	56	5	6	18	63	27	20	24
Upper limb distal muscles								
Wrist flexion	48	16	22	0	63	18	27	28
Wrist extension	51	19	22	9	63	59	23	31
Finger flexion	42	23	59	0	66	29	24	33
Finger extension	48	43	41	18	66	88	36	44
Interossei	42	64	56	9	62	88	39	49
Lower limb proximal muscles								
Hip extension	96	32	3	71	90	33	55	51
Hip abduction	96	37	3	63	89	18	54	50
Hip flexion	90	27	9	56	91	43	55	50
Knee extension	87	13	11	40	91	35	33	39
Knee flexion	77	23	7	60	70	51	56	46
Lower limb distal muscles								
Ankle dorsiflexion	48	64	30	50	40	73	51	50
Ankle plantar flexion	26	41	24	57	17	71	50	40
Toe extension	47	72	50	50	40	68	55	55
Toe flexion	38	61	6	50	0	78	46	41

ALS, amyotrophic lateral sclerosis; BMD, Becker muscular dystrophy; DMD, Duchenne muscular dystrophy; FSHD, facio-scapulo-humeral dystrophy; HN, hereditary neuropathy; LGMD, limb girdle muscular dystrophy; MD, myotonic dystrophy; NMD, neuromuscular disorders; SMA, spinal muscular atrophy.

the literature.[14 22–24] This procedure determines the extent to which a patient with NMD deviates from normal given his/her gender and age for grip strength and given his/her gender, age and height for gait speed. Grip strength and gait speed were considered as significantly impaired when the z score was lower than −2.

The grades of the MMT represent ordinal scores and are separated by unknown distances. Therefore, mathematical operations on such scores can lead to an incorrect interpretation of results.[25] For these reasons, the scores of the MMT were individually analysed. When the MMT score was less than 5 on the MRC scale, this muscle was considered to be weaker than normal.

The relationships between motor impairment and activity limitations were studied in each of the six main diagnostic groups and in the whole sample without diagnosis distinction. A Pearson's correlation coefficient was used to determine the strength of the linear relationship between the z scores of grip strength and of gait speed and the ACTIVLIM measures while a Spearman's correlation coefficient was used for the relationship between the ordinal score of the MMT and the ACTIVLIM measures. Analyses were performed with the SigmaStat software and a p value <0.05 was considered statistically significant.

RESULTS

Motor impairments

The extent of each motor impairment in the six diagnostic groups and in the total sample is shown in table 3 by the proportion of patients that obtained a z score less than −2 on the grip strength test, a z score less than −2 on the gait speed test and a score less than 5 on the MMT. Moreover, as the scores of MMT showed no significant difference between the right and left sides for all muscle groups (Wilcoxon Signed Rank test, $p>0.1$), the values for the right side were arbitrarily chosen for further analyses.

Activity limitations

The activity limitation measures of patients with NMD are presented concisely here because they were described in a previous study.[6] The ACTIVLIM measures were significantly different with regard to type of NMD ($F = 10.55$, $p<0.001$). The post hoc analysis indicates that the patients with proximal NMD (mean −1.1 (SD 3.07) logits in the DMD/BMD/LGMD group and −0.46 (SD 3.43) logits in SMA group) have a lower activity level than the group with other NMD (mean 0.66 (SD 2.35) logits), the FSHD group (mean 1.56 (SD 1.99) logits), the HN group (mean 1.97 (SD 2.1) logits) and the MD group (mean 2.71 (SD 2.1) logits). The MD group also had a significantly higher activity level than the ALS group (mean 0.39 (SD 2.31) logits) and the group with other NMD.

Relationships between motor impairments and activity limitations

The relationships between motor impairments and activity limitations were not different in children with NMD compared with adults with NMD. Therefore, the results for children and adults were pooled.

Figure 1 shows the relationships between gait speed (fig 1A), grip strength (fig 1B) and activity limitations for each of the six main diagnostic groups. Activity limitations were significantly correlated with gait speed for the DMD/BMD/LGMD, HN, MD and ALS groups ($r = 0.61$, 0.63, 0.71 and 0.69, respectively). No relationship between activity limitations and gait speed was

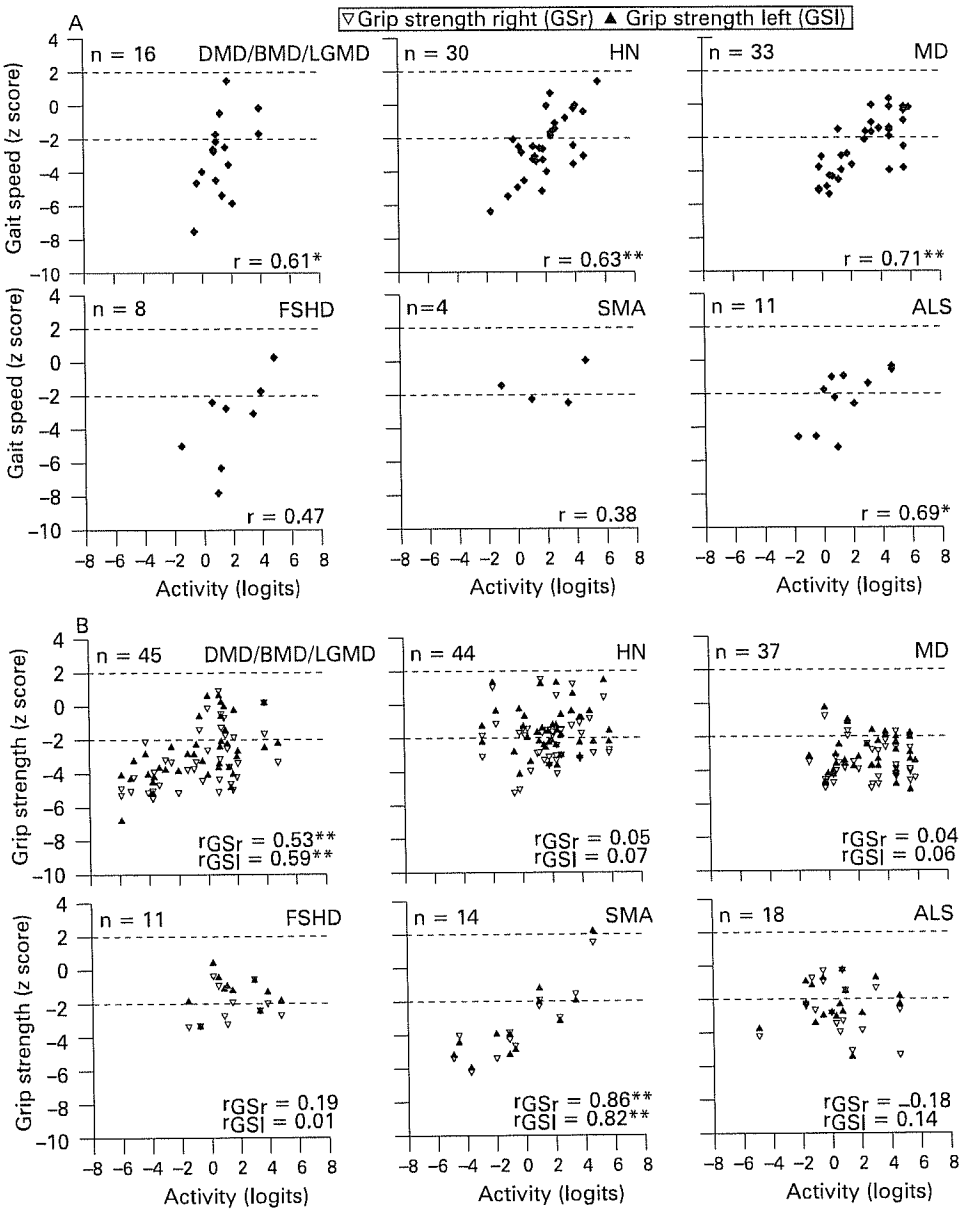

Figure 1 Relationships between activity limitations measured by the ACTIVLIM questionnaire and gait speed (A), grip strength in the right hand (B) and grip strength in the left hand (B) for the six main diagnostic groups (Duchenne, Becker and limb girdle muscular dystrophy (DMD/BMD/LGMD), hereditary neuropathy (HN), myotonic dystrophy (MD), facio-scapulo-humeral muscular dystrophy (FSHD), spinal muscular atrophy (SMA) and amyotrophic lateral sclerosis (ALS)). The Pearson correlation coefficients between activity limitations and gait speed, grip strength in the right hand (r_{GSr}) and grip strength in the left hand (r_{GSl}) are reported in the lower right corner of each figure. The broken lines show a z score of 2 and a z score of −2, the limits between which a patient obtained a z score not significantly different from normal values. *p<0.05, **p<0001.

found in the SMA and FSHD groups. Activity limitations were significantly correlated with grip strength only in the proximal NMD groups with a higher correlation in the SMA group (r = 0.86 and 0.82) than in the DMD/BMD/LGMD group (r = 0.53 and 0.59). No relationship between activity limitations and grip strength was found in the HN, MD, FSHD or ALS groups. Figure 2 shows the relationship between gait speed, grip strength and activity limitations in the whole sample of patients with NMD. Activity limitations were significantly but poorly correlated with grip strength in both hands (r = 0.3 and 0.36) and moderately correlated with gait speed (r = 0.56).

The relationships between the MMT and activity limitations are reported in table 4. The correlations between activity limitations and the MMT of each muscle group largely varied according to the diagnostic groups (range ρ = −0.5 to 0.95 for interossei muscles in the FSHD and SMA groups, respectively). Nevertheless, in the whole sample, activity limitations moderately to poorly correlated with the MMT of each muscle group (range ρ = 0.50 to 0.17 for knee flexors and finger flexors, respectively). The flexor and proximal muscle groups tended to have a stronger relationship with the activity limitations (range ρ = 0.50 to 0.44) than the extensor and distal muscle groups.

DISCUSSION

Relationships between motor impairments and activity limitations assessed by the ACTIVLIM questionnaire were investigated in six main diagnostic groups of NMD and in 245 patients with NMD, all diagnoses taken together.

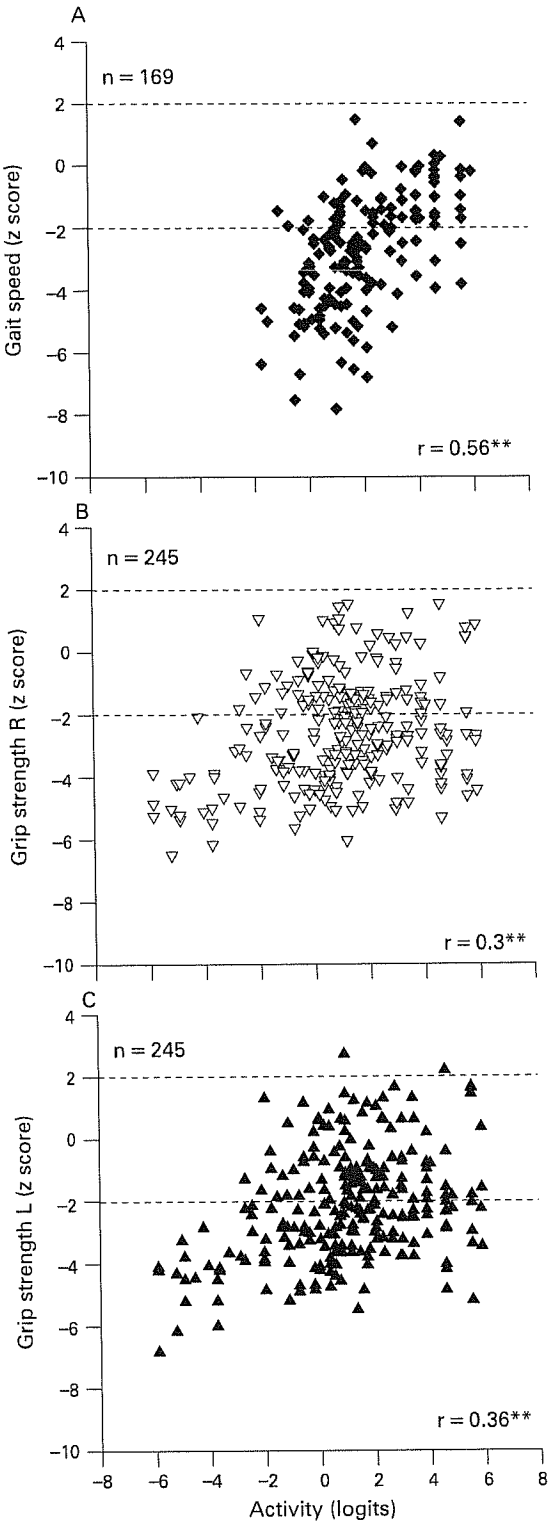

Figure 2 Relationships between activity limitations measured by the ACTIVLIM questionnaire and gait speed (A), grip strength in the right hand (B) and grip strength in the left hand (C) for all patients in the sample. The broken lines show a z score of 2 and a z score of −2, the limits between which a patient obtained a z score not significantly different from normal values. **p<0.001.

The spontaneous gait speed adopted by the patients showed the highest correlation with the ACTIVLIM measures (fig 2A) among the motor impairments assessed. Spontaneous gait speed was considered a measure of locomotor impairment[18] and was calculated from the 10 m walking test. However, this test is, in some respects, applicable to the components "body function and structure" and "activity" of the ICF as it can also reflect difficulties in performing an activity such as walking.[1] This link between the measure of gait speed and the activity limitations measured by ACTIVLIM could explain their good correlation. Moreover, the walking activity is largely represented in the ACTIVLIM measures as five of the 22 items included in the questionnaire are directly related to the walking ability of the patient. Nevertheless, gait speed can give a measure for walking patients alone while the ACTIVLIM measures can also distinguish the activity level of wheelchair bound patients.

Gait speed was moderately correlated with activity limitations in patients with DMD/BMD/LGMD, HN, MD and ALS (r = 0.61 to 0.71 in fig 1A). These correlations may partially be explained by the weakness in the proximal muscles of the lower limbs in patients with DMD/BMD/LGMD and by the weakness in the distal muscles of the lower limbs in patients with HN and ALS. Proportions of patients with MD presenting muscle weakness in the lower limbs (table 4) were, however, quite low. Reduced gait speed and its relationships with activity limitations could eventually be associated with daytime sleepiness, apathy or lack of motivation, which are clinical aspects frequently encountered in patients with MD.[26] No relationship was found in patients with FSHD and SMA, probably because of the small number of walking subjects in each of these diagnostic groups (seven and four, respectively).

Grip strength was moderately correlated with activity limitations in patients with DMD/BMD/LGMD (r = 0.53 and 0.59) and strongly correlated with activity limitations in patients with SMA (r = 0.86 and 0.82), indicating that grip strength could be an indicator of the activity level in patients with DMD/BMD/LGMD and SMA. Nevertheless, a larger sample of patients with SMA is needed to confirm this assumption. On the other hand, no correlation was found between these two variables in patients with HN, MD, FSHD and ALS, indicating that reduced grip strength does not necessarily exclude a high level of activity. The onset of HN, MD and ALS is often characterised by hand and finger muscle weakness[27–29] that can be detected by grip weakness[30] without yet affecting the activity level of these patients.

The disparity of Spearman's correlation coefficients between MMT and activity limitations reflects the discrepancies of the anatomical basis and physiopathology of the NMD. These differences are most obvious between the DMD/BMD/LGMD, HN and MD groups, each of which had almost the same number of patients (n = 45, 44 and 37, respectively) and which therefore allows some comparisons in the values of correlation coefficients. The activity level of patients with DMD/BMD/LGMD seems to be more dependent on muscle weakness than for those with HN or MD. Indeed, unlike the DMD/BMD/LGMD group, muscle weakness and atrophy were not the only clinical signs in these latter groups; sensory loss and feet deformities in HN and myotonia, daytime sleepiness or lack of motivation in MD could also influence the difficulties in performing daily activities, as assessed by the ACTIVLIM questionnaire.[26–28] Very strong correlations between MMT and activity limitations were found in the wrist and hand muscle groups of patients with SMA, in the ankle and toe extensors of patients with FSHD and in the toe flexors in patients with ALS. Nevertheless, caution in the interpretation of these results should be exercised because of the small number of patients in these groups (n = 14, 11 and 18, respectively).

Table 4 Spearman's coefficients of correlation between activity limitations and MMT

	DMD/BMD LGMD (n = 45)	HN (n = 44)	MD (n = 37)	FSHD (n = 11)	SMA (n = 14)	ALS (n = 18)	Total sample (n = 245)
Upper limb proximal muscles							
Shoulder flexion	0.73**	0.27	0.23	0.59	0.45	0.51*	0.47**
Shoulder abduction	0.61**	0.20	0.30	0.66*	0.25	0.38	0.43**
Elbow flexion	0.75**	0.25	0.24	0.53	0.59*	0.10	0.48**
Elbow extension	0.68**	0.26	0.39*	0.40	0.29	0.05	0.40**
Upper limb distal muscles							
Wrist flexion	0.43*	−0.16	0.42**	0.01	0.88**	0.18	0.27**
Wrist extension	0.45*	−0.02	0.34*	0.10	0.88**	0.06	0.34**
Finger flexion	0.62**	0.03	0.35*	0.01	0.93**	−0.02	0.17*
Finger extension	0.73**	−0.07	0.53**	0.35	0.94**	0.10	0.31**
Interossei	0.63**	0.21	0.46**	−0.50	0.95**	0.20	0.25**
Lower limb proximal muscles							
Hip extension	0.59**	0.04	0.19	0.50	0.73*	0.37	0.39**
Hip abduction	0.58**	0.25	0.01	0.28	0.56	0.39	0.46**
Hip flexion	0.71**	−0.03	0.16	0.47	0.46	0.45	0.44**
Knee extension	0.68**	0.02	0.35*	0.69*	0.58	0.42	0.45**
Knee flexion	0.72**	0.45*	0.18	0.59	0.73*	0.66*	0.50**
Lower limb distal muscles							
Ankle dorsiflexion	0.56**	0.28	0.40*	0.84**	0.65*	0.36	0.34**
Ankle plantar flexion	0.75**	0.28	0.51**	0.39	0.40	0.56	0.40**
Toe extension	0.69**	0.20	0.39*	0.77**	−0.03	0.59*	0.35**
Toe flexion	0.83**	0.36	0.14	0.01	0.32	0.88**	0.49**

ALS, amyotrophic lateral sclerosis; BMD, Becker muscular dystrophy; DMD, Duchenne muscular dystrophy; FSHD, facio-scapulo-humeral dystrophy; HN, hereditary neuropathy; LGMD, limb girdle muscular dystrophy; MD, myotonic dystrophy; MMT, Manual Muscle Testing; SMA, spinal muscular atrophy.
*0.05>p>0.01, **p<0.01.

Considering the whole sample, the relationships between activity limitations and MMT were poor to moderate, indicating that high levels of activity do not necessarily require full muscle strength. Indeed, the MMT is an analytical measure while the achievement of daily activities is a combination of movements involving several muscle groups. If activity limitations are now correlated with a global MMT sum score or with an arm or a leg sum score to investigate the strength contribution of all muscle groups on activity limitations, Spearman's correlation coefficients were equal to 0.55, 0.45 and 0.55, respectively. These correlations were not much higher than the reported correlations using the individual scores of the MMT. However, the results of the MMT sum scores should be interpreted with caution because of the ordinal nature of the MMT scores and the incorrect use of mathematical operations on such scores.[25] The use of the MMT grades could also induce some measurement errors in the calculation of the correlation coefficients because grade 4 of the MMT covers a wide range of strength[31] and therefore may not be precise enough to discriminate between different activity levels.

Patients with muscle weakness can also develop compensatory strategies that allow them to complete daily activities and, consequently, to have a higher activity measure. Moreover, other impairments such as fatigue, pain, contractures, respiratory or sensory impairments could contribute to activity limitations in patients with NMD.[27 32-35] Finally, personal contextual factors (eg, motivation, cognition) and environmental factors (eg, financial support, health services)[1] could facilitate or hinder the achievement of daily activities and thus should be considered in rehabilitation processes.[36 37] However, future studies are needed to identify which contextual factors contribute to the achievement of daily activities. Furthermore, the study of participation restrictions in patients with NMD and of their relationships with motor impairments and activity limitations is also essential to have an overall view of interactions between each component of the ICF.

Despite the moderate correlations between muscle strength measures and activity limitations, the values of the correlation coefficients in the whole sample were consistent with the daily activities included in the ACTIVLIM.[8] On the one hand, the correlations of the MMT of the flexors and proximal muscle groups with the ACTIVLIM measure emphasise the need to use these muscles to achieve the 22 daily activities of the ACTIVLIM. For example, washing one's face and putting on a T-shirt require functional shoulder and elbow flexors to be easily performed. Similarly, when walking upstairs or stepping out of a bath tub, functional hip and knee flexors are needed to lift the legs. On the other hand, the items of the ACTIVLIM do not include activities requiring hand and finger strength, with the result that grip strength measures and the MMT scores of the hands and wrists have the lowest correlation with the ACTIVLIM measures.

The main limitation of this study is the lack of regression analysis, allowing motor impairments to be combined in order to predict the highest proportion of the variance in measures of activity limitations. Multiple linear regression analysis cannot be performed in this study because of the ordinal nature of the MMT data. To overcome this problem, non-parametric regression could be used but a larger sample would be necessary to obtain conclusive results. Measuring muscle strength with a quantitative technique, such as quantitative muscle testing, could be another method allowing multiple linear regression to be performed. Indeed, quantitative muscle testing has the advantage of being a linear measure but it is also recognised as a more objective and sensitive measure of muscle strength than the MMT scores.[31 38] These further analyses could be interesting in the whole sample but also in the different diagnostic groups.

Although all motor impairments were significantly related to a decrease in activity level, their relationships were poor to moderate with measures of activity limitations. This does not mean that clinical tests such as the MMT, grip strength or 10 m walking test are useless, but they cannot precisely predict what patients can perform as daily activities in their usual environment. For this reason, activity limitations should be measured separately. This result supports the theoretical standpoint of the ICF that motor impairments and activity level are not related in a predictably straightforward way.[1] Although this relationship has rarely been investigated in patients with neuromuscular disorders, our observations confirm other studies.[2 4 5] In patients with immune mediated polyneuropathies, 64% of the variance of the disabilities was explained by impairment measures.[2] In patients with DMD, correlations between strength measures and functional ability assessed by the Functional Independence Measure or the Barthel Index ranged from 0.39 to 0.56.[4 5] Therefore, the reduction in motor impairments could not directly result in a corresponding higher activity level. Consequently, interventions focused on daily activities should continue to be the most important aim in patient rehabilitation in order to preserve the patient's independence for as long as possible.[39] An ideal intervention should always endeavour to coordinate physical therapy for its key role in preventing a decrease in muscle strength and joint immobility and occupational therapy for its key role in the management of meaningful daily activities. Teaching patients how to optimise their motor function should be an important part of rehabilitation as it can help patients developing adaptive strategies to compensate for their motor impairments.[40]

The present study has stressed the importance of treating and measuring activity level independently as it is not simply the integration of motor function in daily activities. It does not mean that interventions intended to reduce motor impairments are useless but rather are complementary to those intended to improve patients' activity levels.

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Note: The ACTIVLIM questionnaire and its administration instructions can be downloaded from www.rehab-scales.org in English, French and Dutch. The website also allows total raw scores for the ACTIVLIM questionnaire to be converted into a linear measure of activity limitations, according to the Rasch model.

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Competing interests: None.

Ethics approval: The study was approved by the medical ethics committees of the Université catholique de Louvain and the Katholieke Universiteit Leuven.

REFERENCES

1. **World Health Organization.** *The International Classification of Functioning, Disability and Health-ICF.* Geneva: WHO, 2001.
2. **Merkies IS,** Schmitz PI, van der Meche FG, *et al.* Connecting impairment, disability, and handicap in immune mediated polyneuropathies. *J Neurol Neurosurg Psychiatry* 2003;**74**:99–104.
3. **Arnould C,** Penta M, Thonnard JL. Hand impairments and their relationship with manual ability in children with cerebral palsy. *J Rehabil Med* 2007;**39**:708–14.
4. **Uchikawa K,** Liu M, Hanayama K, *et al.* Functional status and muscle strength in people with Duchenne muscular dystrophy living in the community. *J Rehabil Med* 2004;**36**:124–9.
5. **Nair KP,** Vasanth A, Gourie-Devi M, *et al.* Disabilities in children with Duchenne muscular dystrophy: a profile. *J Rehabil Med* 2001;**33**:147–9.
6. **McDonald CM.** Physical activity, health impairments, and disability in neuromuscular disease. *Am J Phys Med Rehabil* 2002;**81**:S108–20.
7. **Hogrel JY,** Ollivier G, Desnuelle C. Manual and quantitative muscle testing in neuromuscular disorders. How to assess the consistency of strength measurements in clinical trials? *Rev Neurol (Paris)* 2006;**162**:427–36.
8. **Vandervelde L,** Van den Bergh PY, Goemans N, *et al.* ACTIVLIM: a Rasch-built measure of activity limitations in children and adults with neuromuscular disorders. *Neuromuscul Disord* 2007;**17**:459–69.
9. **Hornyak JEt, Pangilinan PH Jr.** Rehabilitation of children and adults who have neuromuscular diseases. *Phys Med Rehabil Clin N Am* 2007;**18**:883–97.
10. **Spiegel JS,** Paulus HE, Ward NB, *et al.* What are we measuring? An examination of walk time and grip strength. *J Rheumatol* 1987;**14**:80–6.
11. **Thyberg I,** Hass UA, Nordenskiold U, *et al.* Activity limitation in rheumatoid arthritis correlates with reduced grip force regardless of sex: the Swedish TIRA project. *Arthritis Rheum* 2005;**53**:886–96.
12. **Giampaoli S,** Ferrucci L, Cecchi F, *et al.* Hand-grip strength predicts incident disability in non-disabled older men. *Age Ageing* 1999;**28**:283–8.
13. **Rantanen T,** Guralnik JM, Foley D, *et al.* Midlife hand grip strength as a predictor of old age disability. *JAMA* 1999;**281**:558–60.
14. **Mathiowetz V,** Kashman N, Volland G, *et al.* Grip and pinch strength: normative data for adults. *Arch Phys Med Rehabil* 1985;**66**:69–74.
15. **Haigh R,** Tennant A, Biering-Sorensen F, *et al.* The use of outcome measures in physical medicine and rehabilitation within Europe. *J Rehabil Med* 2001;**33**:273–8.
16. **Council MR.** *Aids to the examination of the peripheral nervous system. Memorandum 45.* London: Pendragon House, 1976.
17. **Kendall F,** Mc Creary E. *Muscles: testing and function.* Baltimore: Lippincott Williams & Wilkins, 1993.
18. **Bernardi M,** Macaluso A, Sproviero E, *et al.* Cost of walking and locomotor impairment. *J Electromyogr Kinesiol* 1999;**9**:149–57.
19. **Goldfarb BJ,** Simon SR. Gait patterns in patients with amyotrophic lateral sclerosis. *Arch Phys Med Rehabil* 1984;**65**:61–5.
20. **Solari A,** Laura M, Salsano E, *et al.* Reliability of clinical outcome measures in Charcot–Marie–Tooth disease. *Neuromuscul Disord* 2008;**18**:19–26.
21. **Rasch G.** *Probabilistic models for some intelligence and attainment tests.* Chicago: MESA Press, 1980.
22. **Bohannon RW.** Comfortable and maximum walking speed of adults aged 20–79 years: reference values and determinants. *Age Ageing* 1997;**26**:15–19.
23. **Mathiowetz V,** Wiemer DM, Federman SM. Grip and pinch strength: norms for 6- to 19-year-olds. *Am J Occup Ther* 1986;**40**:705–11.
24. **Wheelwright EF,** Minns RA, Law HT, *et al.* Temporal and spatial parameters of gait in children. I: Normal control data. *Dev Med Child Neurol* 1993;**35**:102–13.
25. **Merbitz C,** Morris J, Grip JC. Ordinal scales and foundations of misinference. *Arch Phys Med Rehabil* 1989;**70**:308–12.
26. **van der Werf S,** Kalkman J, Bleijenberg G, *et al.* The relation between daytime sleepiness, fatigue, and reduced motivation in patients with adult onset myotonic dystrophy. *J Neurol Neurosurg Psychiatry* 2003;**74**:138–9.
27. **Pareyson D.** Differential diagnosis of Charcot–Marie–Tooth disease and related neuropathies. *Neurol Sci* 2004;**25**:72–82.
28. **Schara U,** Schoser BG. Myotonic dystrophies type 1 and 2: a summary on current aspects. *Semin Pediatr Neurol* 2006;**13**:71–9.
29. **Mitchell JD,** Borasio GD. Amyotrophic lateral sclerosis. *Lancet* 2007;**369**:2031–41.
30. **Kozin SH,** Porter S, Clark P, *et al.* The contribution of the intrinsic muscles to grip and pinch strength. *J Hand Surg (Am)* 1999;**24**:64–72.
31. **Mendell JR,** Florence J. Manual muscle testing. *Muscle Nerve* 1990;**13**(Suppl):S16–20.
32. **Johnson ER,** Fowler WM Jr, Lieberman JS. Contractures in neuromuscular disease. *Arch Phys Med Rehabil* 1992;**73**:807–10.
33. **Tiffreau V,** Viet G, Thevenon A. Pain and neuromuscular disease: the results of a survey. *Am J Phys Med Rehabil* 2006;**85**:756–66.
34. **McDonald CM,** Abresch RT, Carter GT, *et al.* Profiles of neuromuscular diseases. Duchenne muscular dystrophy. *Am J Phys Med Rehabil* 1995;**74**:S70–92.
35. **Feasson L,** Camdessanche JP, El Mandhi L, *et al.* Fatigue and neuromuscular diseases. *Ann Readapt Med Phys* 2006;**49**:289–300, 75–84.
36. **Law M.** Evaluating activities of daily living: directions for the future. *Am J Occup Ther* 1993;**47**:233–7.
37. **Schneidert M,** Hurst R, Miller J, *et al.* The role of environment in the International Classification of Functioning, Disability and Health (ICF). *Disabil Rehabil* 2003;**25**:588–95.
38. **Aitkens S,** Lord J, Bernauer E, *et al.* Relationship of manual muscle testing to objective strength measurements. *Muscle Nerve* 1989;**12**:173–7.
39. **Fowler WM Jr.** Rehabilitation management of muscular dystrophy and related disorders: II. Comprehensive care. *Arch Phys Med Rehabil* 1982;**63**:322–8.
40. **Kakulas BA.** Problems and solutions in the rehabilitation of patients with progressive muscular dystrophy. *Scand J Rehabil Med Suppl* 1999;**39**:23–37.