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ORIGINAL REPORT

LIMITED UPPER LIMB FUNCTIONING HAS IMPACT ON RESTRICTIONS IN PARTICIPATION AND AUTONOMY OF PATIENTS WITH HEREDITARY MOTOR AND SENSORY NEUROPATHY 1A

Annemieke J. Videler, MSc, PT[1], Anita Beelen, PhD[1], Ivo N. van Schaik, MD[2], Marianne de Visser, MD[2] and Frans Nollet, MD[1]

From the [1]Department of Rehabilitation and [2]Department of Neurology Academic Medical Center, University of Amsterdam, Amsterdam, The Netherlands

Objective: **To evaluate upper limb functioning, restrictions on participation and the independent contribution of upper and lower limb disability to participation in hereditary motor and sensory neuropathy 1a.**
Design: **Descriptive cross-sectional study.**
Subjects: **Forty-nine patients with hereditary motor and sensory neuropathy 1a.**
Methods: **Perceived upper limb functioning was evaluated using the Michigan Hand Outcomes Questionnaire and participation restrictions with the Impact on Participation and Autonomy Questionnaire. Upper and lower limb domains of Guy's Neurological Disability Scale were used to determine their impact on participation restrictions.**
Results: **Limitations in upper limb functioning were perceived by 98% of the patients. Median scores ranged between 70 points for overall hand function and 100 points for aesthetics (scale 0–100). Patients were least satisfied with dominant hand performance. Most patients (46–78%) reported their participation to be sufficient. Restrictions were reported in the domains work, family roles, and autonomy outdoors. Minor problems with restricted participation were indicated by 22–55%, severe problems by 2–12%. Upper limb functioning correlated significantly with all participation subscales. Upper limb disability was independently associated with participation restrictions, whereas lower limb disability was not.**
Conclusion: **Limitations in upper limb functioning were perceived by the majority of patients with hereditary motor and sensory neuropathy 1a and strongly related to restricted participation.**

Key words: hereditary motor and sensory neuropathies, Charcot-Marie-Tooth disease, upper extremity, hand, disability evaluation.

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Correspondence address: Annemieke J. Videler, Department of Rehabilitation, A01-415, Academic Medical Center, University of Amsterdam, PO Box 22660, NL-1100 DD Amsterdam, The Netherlands. E-mail: a.j.videler@amc.uva.nl

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INTRODUCTION

Hereditary motor and sensory neuropathy (HMSN), also known as Charcot-Marie-Tooth disease (CMT), is the most common inherited neuropathy, affecting approximately one in every 2500 individuals (1). Demyelinating HMSN1a, the most prevalent subtype of HMSN, is characterized by slowly progressive distal muscle weakness and wasting and by sensory loss. In the upper limbs the intrinsic hand muscles are primarily affected, often manifesting as a clawing of the fingers and impaired hand function (2–5).

The progression of HMSN1a cannot currently be influenced. The rehabilitation of these patients therefore aims to reduce the limitations in activities and optimize participation in society (6). A previous pilot study of 20 patients with HMSN type I and II showed that 25% of the patients perceived major limitations in hand function (7). More recently, we studied the ability of HMSN1a patients to execute manual tasks and showed that impaired manual dexterity is common, especially for activities requiring finger grips (8). Although these latter results provided information on the capacity of HMSN1a patients to perform manual tasks, they did not provide insight into patients' perception of functioning. Participation, defined by the International Classification of Functioning, Disability and Health (ICF) as the social involvement in a life situation, is a concept that addresses the need to assess patients' own perceptions of their life situation (6). Quality of life in HMSN has attracted interest recently (9–12), but to our knowledge no reports have been published on either perceived upper limb functioning or patient participation in HMSN1a. Lack of insight into the perceived limitations and restrictions in participation hampers practitioners in informing patients about the functional and societal consequences of their disease and in determining effective rehabilitation programmes.

The aim of the present study was to evaluate perceived upper limb functioning and restrictions in participation in patients with HMSN1a. In HMSN1a both lower and upper limbs are involved, with the lower limbs becoming affected earlier and more severely. Therefore, the independent contribution of upper and lower limb functioning to participation restrictions was also studied.

METHODS

Participants

The study sample consisted of 49 HMSN1a patients who had already been described in detail previously (8). All patients met the following inclusion criteria: between 18 and 70 years of age and diagnosis

confirmed by a duplication on chromosome 17p11.2–p12. Patients were excluded if they had difficulties comprehending or reading the Dutch language and if their medical history included stroke, plexopathy, radiculopathy, upper limb pathology (traumatic, degenerative, idiopathic), upper limb surgery or a psychiatric disorder.

Written informed consent was obtained from all patients in accordance with the protocol approved by the university hospital medical ethics committee.

Assessments

Perceived upper limb functioning was evaluated using the Dutch language version of the Michigan Hand Outcomes Questionnaire (MHQ-DLV). The MHQ is a 57-item hand-specific outcome questionnaire designed specifically for the assessment of perceived physical functioning of patients with chronic hand conditions. Furthermore, it provides separate domain scores relevant for HMSN1a patients. This self-administered questionnaire contains 6 domains: 1) overall hand function, 2) activities of daily living, 3) pain, 4) work performance, 5) aesthetics, or the appearance of the hand, and 6) patient satisfaction with hand function. Domains 1, 2, 5, and 6 assess each hand separately. In general, the mean time required to complete the questionnaire is 10 min (13). Raw scores for each scale can be converted to a sum score ranging from 0 to 100; high scores denote better hand performance and less pain. The MHQ has been proven to be reliable, valid and responsive to change in patients with chronic hand disorders (13–15). Though not specifically validated for HMSN, it has been used in various hand pathology studies, such as carpal tunnel syndrome, Dupuytren's disease and full-thickness hand burns (14, 16–18).

The Impact on Participation and Autonomy Questionnaire (IPA) was used to assess restrictions in participation (19, 20). We have chosen this generic self-administered questionnaire as it focuses on patient's self perception of participation. The IPA measures 2 aspects of participation. Firstly, the IPA quantifies restrictions in 5 domains reflecting different life situations: 1) autonomy indoors, 2) autonomy outdoors, 3) family role, 4) social relationships, and 5) work and education. Each domain is graded on a 5-point scale, with responses ranging from 0 (very good) to 4 (very poor). A mean score was calculated for each domain; higher mean scores denote more restrictions on participation in the specific domain. Secondly, the IPA examines 9 items (mobility, self-care, family role, finances, leisure, social relations, helping and supporting others, occupation, and education) to determine the extent to which patients perceive their restrictions on participation as problematic. This second aspect was graded on a 3-point scale, ranging from 0 (no problem) to 2 (severe problem). The IPA is considered to be reliable and has been validated in patients with neuromuscular disease, spinal cord injury, stroke, rheumatoid arthritis, traumatic hand injury and fibromyalgia (19–22).

We used the upper and lower limb domains of the Guy's Neurological Disability Scale (GNDS) to assess the impact of upper and lower limb disabilities on participation restrictions (23). The GNDS was developed as a measure of disability in multiple sclerosis and found to be reliable, responsive, and valid (23, 24). This face-to-face questionnaire has also been applied to inflammatory neuropathies and HMSN studies (25, 26) and was chosen as it allows for a standardized and similar evaluation of both lower and upper limb disabilities. Scores range between 0 and 5 (most severe problem in that area) and are graded for the upper limb and lower limb as: 0 = no problems, 1 = problems in one or both arms, not affecting activities such as dressing, washing or brushing hair, turning a key, eating and doing up zips or buttons; walking is affected, but walks independently; 2 = problems in one or both arms affecting some, but not preventing any, of the activities listed; mostly uses unilateral support to walk outdoors; 3 = problems in one or both arms, affecting all or preventing one or 2 of the activities listed; mostly uses bilateral support to walk outdoors; 4 = problems in one or both arms preventing 3 of the activities listed; mostly uses wheelchair to travel outdoors, but able to stand and walk a few steps and 5 = unable to use either arm for any purposeful movements; restricted to wheelchair, unable to walk a few steps.

Data concerning patient characteristics and MHQ and IPA data were collected by one examiner, as part of a larger descriptive cross-sectional study on the determinants of manual dexterity in HMSN1a. The GNDS was scored by a neurologist.

Statistical analysis

The characteristics of the study group were summarized by descriptive statistics.

Mean and standard deviation (SD) or median and (25th and 75th percentile) scores were calculated for all domains of the MHQ, GNDS, and subscales of the IPA. Differences between dominant and non-dominant MHQ scores were compared by paired sample *t*-tests or Wilcoxon signed-rank tests. Associations between perceived upper limb functioning determined by the MHQ, participation scores determined by the IPA, and age were investigated using Pearson product moment correlation coefficients (r) or Spearman rank correlation coefficients (r_s) depending on the distribution of the data.

The independent contribution of upper and lower limb disabilities to perceived restrictions on the different domains of participation, as determined by the GNDS scores, was explored using multiple linear regression analysis. Residual analysis was performed to search for violations of necessary assumptions in the multiple regression in terms of linearity, equality of variance, independence of error, normality, and influential data points (Cook's distances). Statistical significance was set at $p < 0.05$. All statistical analyses were performed using SPSS 16.0.2 for Windows.

RESULTS

From the 63 DNA confirmed HMSN1a patients, known at the departments of neurology and rehabilitation medicine of our university hospital, 53 agreed to participate. Four patients were excluded: 3 with co-morbidity (Dupuytren's disease, recent shoulder surgery, psychiatric disorder) and one with alcohol abuse in the history. The characteristics of the final study sample ($n = 49$, 28 women and 21 men) are shown in Table I. Affected hand function was perceived by the majority of participants. Disease duration, duration of hand involvement, and working status varied greatly.

Perceived upper limb functioning

Ninety-eight percent of the HMSN1a patients perceived limitations in upper limb functioning (MHQ sum score < 100). The domain and sum scores for the MHQ are shown in Table II. Median scores are given, as the distribution of

Table I. *Characteristics of 49 patients with hereditary motor and sensory neuropathy 1a (HMSN)*

Characteristics	
Sex, male/female, *n*	21/28
Age, years, mean (range)	46.8 (21–69)
Hand dominance, right/left, *n*	48/1
Disease duration*, years, mean (range)	31.5 (2.7–60.1)
Hand involvement, yes/no/missing, *n*	37/11/1
Duration of hand involvement†, years, mean (range)	14.2 (1.6–54.8)
Working status, yes/no/not applicable, *n*	32/16/1
Disability benefit, yes/no/not applicable, *n*	22/25/2

*Time since first symptoms of HMSN.
†Time since first hand symptoms, if applicable ($n = 37$).

Table II. *Michigan Hand Outcomes Questionnaire (MHQ) domain and sum scores (n = 49)*

Domain		Median (P25/P75)	Range
Overall hand function	D	70.0 (55.0/75.0)	35–100
	ND	70.0 (65.0/75.0)	35–100
ADL	D	85.4 (72.1/95.7)	16–100
	ND	85.4 (73.6/94.5)	11–100
Work		80.0 (52.5/100)	0–100
Pain*		90.0 (60.0/100)	5–100
Aesthetics	D	100.0 (81.3/100)	31–100
	ND	93.8 (87.5/100)	38–100
Satisfaction†	D	75.0 (45.8/93.8)	17–100
	ND	83.3 (54.2/100)	13–100
Sum score†	D	80.3 (63.8/92.5)	21–100
	ND	82.4 (66.3/91.7)	21.2–99.3

*Recorded pain score (100 = no pain), †Significant differences between dominant (D) and non-dominant (ND) hand scores ($p < 0.01$).
Median scores, 25th and 75th percentile (P25 and P75) scores, and minimum and maximum scores are given for all domains of the MHQ; high scores denote better hand performance. ADL: activities of daily living.

MHQ scores appeared to be skewed. Most limitations were indicated in the domains of overall hand function, work, and satisfaction with hand function. Median scores ranged between 70 points for overall hand function and 100 for aesthetics. High MHQ scores were found for 2 domains: aesthetics and pain. The MHQ sum score for the dominant hand was significantly lower than the non-dominant hand ($p < 0.001$, Wilcoxon signed-rank test). At the domain level, a significant difference between both hands was only found for one domain, satisfaction ($p < 0.001$). A significant negative association was found between the MHQ sum scores for the dominant hand and age (Spearman's rank correlation coefficient: –0.31, $p < 0.05$).

Perceived restrictions on participation and autonomy

The standardized median (P25; P75) sum scores for all IPA subscales are given in Table III. A wide range of scores was found. For example, in the work and education domain the scores ranged from 0 (no restrictions) to 4, which indicates very poor participation and autonomy. The extent to which patients quantified their participation is also shown in Table III. Overall, the majority of patients (46–78%) reported their participation to be very good or good. Fair participation was perceived by more than 25% of the patients for all of the IPA subscales, with the exception of autonomy indoors. In work and education, more than one-third of the patients experienced fair participation and autonomy. Reports of insufficient participation, either poor or very poor, was indicated in the domains of getting or keeping work, doing preferred work, social contacts, leisure time, moving freely indoors and outdoors, and performing the role, tasks, and responsibilities within the family.

Table IV shows the percentage of patients reporting problems in 9 aspects of participation. A large proportion (22–55%) of the patients perceived minor problems. Severe problems with restricted participation were indicated by a small group (2–12%) of the HMSN1a patients, particularly with items addressing their family role, leisure, and (voluntary) occupation.

All correlations between MHQ sum scores and the IPA subscale scores were significant ($p < 0.01$; Table V). No significant association was found between the IPA domain scores and age.

Impact of upper and lower limb functioning on participation

The median (P25; P75, range) GNDS score was 2 (1; 2, 0–3) for the upper limb and 3 (2; 3, 0–4) for the lower limb. For one patient no GNDS scores were obtained due to a failure to visit the participating neurologist. The independent contributions of upper and lower limb disabilities to perceived restrictions on

Table III. *Standardized scores of the Impact on Participation and Autonomy Questionnaire – Perceived restrictions on participation (n = 49)*

		Standardized sum scores Median (P25/P75) [Range]	Perceived participation *n* (%)		
Subscale	Content		Very good & Good	Fair	Poor & Very poor
Autonomy indoors	Chances of looking after yourself the way you want (washing, dressing, going to bed, eating, and drinking), moving freely indoors.	0.7 (0.0/1.0) [0–2.9]	38 (77.6)	9 (18.4)	2 (4.1)
Family role	Performing your role, tasks, and responsibilities within the family, doing tasks around the house and garden, choosing how to spend your money.	1.1 (0.7/2.0) [0–3.4]	24 (49.0)	14 (28.6)	3 (6.1)
Autonomy outdoors	Activities outdoors, such as frequency of social contacts, possibilities to spend leisure time, and to get around outdoors when and where you want, leading the life you want.	1.0 (0.6/1.8) [0–3.2]	25 (51)	17 (34.7)	7 (14.3)
Social life and relationships	Quality of social life and relationships, communication, respect, and intimacy. Helping and supporting other people the way you want.	0.7 (0.4/1.1) [0–2.0]	37 (75.5)	12 (24.5)	–
Work and education*	Paid and voluntary work, education and training.	1.2 (0.8/1.8) [0–4.0]	17 (45.9)	14 (37.8)	6 (16.2)

*Twelve patients reported that the item was not relevant to them and, according to the instructions in the questionnaire, omitted this subscale.
Standardized median sum scores range from 0 to 4; 0 = very good participation and autonomy, 1 = good, 2 = fair, 3 = poor, and 4 = very poor.
P25: 25th percentile; P75: 75th percentile.

Table IV. *Results of the Impact on Participation and Autonomy Questionnaire (IPA) – Problem experience (n = 49)*

Items on problem experience	No problems (%)	Minor problems (%)	Severe problems (%)
Mobility	41	55	4
Self-care	67	31	2
Family role	43	45	12
Finances	69	27	4
Leisure	41	51	8
Social relations	61	33	6
Helping and supporting	55	41	4
(Voluntary) occupation*	31	41	8
Education†	29	22	4

Values are reported as the percentage of patients that perceived their restrictions on participation as problematic. *Ten and †22 patients, respectively, indicated that these IPA items were irrelevant and, in accordance with the instructions of the IPA, omitted them.

the different domains of participation are shown in Table V. Upper limb disability, as determined by the GNDS, independently contributed to the IPA subscales for indoors, family role, and work and education. Lower limb disability did not significantly contribute to either of the IPA subscales.

DISCUSSION

The majority of the HMSN1a patients perceived limited upper limb functioning and mild restrictions on participation. Patients were satisfied with the appearance of their hands and rarely experienced pain. The dominant hand was reported as being the most limited and patients were subsequently least satisfied with dominant hand functioning; a finding that can be explained by higher expectations and demands for the dominant hand. Restrictions on participation were perceived particularly in the domains of work, family role, and autonomy outdoors. Although most HMSN1a patients found their participation to be sufficient in the domains addressed, a limited group (2–12%) indicated severe problems with restrictions on participation.

Table V. *Univariate and multivariate associations between upper and lower limb functioning and restrictions on participation*

Univariate association		Multivariate association				
IPA subscale	MHQ sum score	GNDS domain scores	B	Std Error	*p*	Adjusted R^2
IPA indoors	–0.67**	Upper limb	0.24	0.12	0.045*	0.13
		Lower limb	0.19	0.11	0.11	
IPA family role	–0.72**	Upper limb	0.41	0.15	0.01*	0.23
		Lower limb	0.19	0.14	0.20	
IPA outdoors	–0.66**	Upper limb	0.26	0.14	0.06	0.19
		Lower limb	0.23	0.13	0.08	
IPA social relations	–0.49**	Upper limb	0.13	0.09	0.14	0.04
		Lower limb	0.03	0.09	0.69	
IPA work	–0.79**	Upper limb	0.41	0.20	0.049*	0.14
		Lower limb	0.11	0.19	0.58	

Univariate association: Spearman's rank correlation coefficients between perceived upper limb function (Michigan Hand Outcomes Questionnaire (MHQ)) and restrictions on participation (Impact on Participation and Autonomy Questionnaire (IPA); $n = 49$) are given.

Multivariate association: the association between upper- and lower limb disabilities (Guy's Neurological Disability Scale (GNDS) domain scores) and IPA domain scores are based on multiple linear regression analyses ($n = 48$). Analysis of residuals did not show violations of necessary assumptions in multiple regression in terms of linearity, equality of variance, independence of error and normality. R^2 is the percentage of the total variation of the dependent variable score (restrictions in participation) that is explained by the independent variables (upper and lower limb disability scores). Each percentage is adjusted for the variables that are already included in the model. Adjusted R^2 is adjusted for the number of independent variables in the model. *$p < 0.05$; **$p < 0.01$.

The MHQ has been found to be reliable and reproducible in patients with various wrist and hand disorders (13, 14, 18), but in HMSN1a the psychometric properties of this questionnaire still needs to be investigated. Compared with the MHQ scores reported for other pathological conditions of the hand, such as full-thickness hand burns (mean 63 (standard deviation (SD) 23)), Dupuytren's contracture (mean 74.7 (SD 12.8)), other peripheral nerve (median 59.9), and wrist disorders (median 83.4), our results indicate that HMSN1a patients perceived, on average, moderate restrictions, though ranging from severe to mild (16–18).

Overall, the degree of perceived restrictions in participation of this cohort of HMSN1a patients was comparable to those of patients with other chronic diseases, such as multiple sclerosis, rheumatoid arthritis, and spinal cord injury (21). In the subscale family role, for example, the median standardized IPA score ranged from 1.14 to 1.6 in the other conditions compared with 1.3 found in the present study. Nevertheless, the kind of perceived restrictions will evidently differ between diseases, as the distribution of impairments is disease specific.

HMSN1a is a chronic disease in which patients have time to adapt to the increasing impairment and limitations in hand functioning. This adaptation to disability and poor health is known as "response-shift" (27) and may contribute to the mildness of the perceived limitations in upper limb function and restrictions on participation and autonomy.

The patients' perceived upper limb functioning appeared to be strongly related to the perceived restrictions on participation and autonomy, especially in the domains of work and family role. These domains contain, in particular, upper limb activities, for example doing work the way one prefers, minor and heavy housework, and repair activities.

Both the lower and upper limbs are involved in HMSN1a, with the lower limbs affected more severely. This was reflected by the median GNDS scores we found in this HMSN1a cohort, indicating that usually bilateral support is needed (2 canes or crutches, frame, or 2 arms) to walk outdoors (lower limb score 3) and that the problems in one or both arms affect but not prevent activities such as dressing, washing or brushing hair, turning a key, eating and doing up zips (upper limb score 2). Surprisingly, the GNDS lower limb scores did not contribute significantly to either of the IPA subscales, although the association between lower limb disability and autonomy outdoors almost reached statistical significance. Upper limb disability contributed independently to the restrictions in participation and autonomy, but the explained variances found were low (< 23%). This may be explained by the fact that the GNDS is an

ordinal 6-step scale, making it insensitive to small differences in disability. As the GNDS evaluates the upper and lower limbs in a similar way, this cannot explain the finding that upper limb disability contributed independently to restrictions in participation and lower limb disability did not. Although we do believe that upper limb function has major impact on participation in HMSN1a, and that these are not spurious findings, further study is needed in this field.

This study is the first to evaluate perceived upper limb functioning and restrictions on participation in a large sample of DNA-confirmed type 1a HMSN patients. Nevertheless, it has a few weaknesses. Firstly, there are no questionnaires available that are specifically designed for HMSN1a. Therefore, the questionnaires used in this study have been chosen based on clinical experience, but have not been validated for patients with HMSN1a. Secondly, the study focused on the patients' perception and used self-assessment questionnaires. As in any study based on self-assessment, the patients' own perception might be influenced by factors such as the attention provided by the study, the presence of an investigator, or the questionnaire itself and, therefore, only approaches reality. Thirdly, the study population was not a random sample of HMSN1a patients. Some selection bias may have occurred because patients with advanced upper limb involvement could be more willing to participate than those who are less impaired. However, it is reasonable to believe that the results provide a good indication of perceived upper limb functioning and participation in HMSN1a because a high percentage (78%) of the known patients in our departments participated, selection was based only on diagnosis, and 22% of our study sample did not indicate upper limb involvement.

In conclusion, this study shows that upper limb functioning in patients with HMSN1a has a clear impact on restrictions on participation and autonomy. These results may support health professionals in offering tailor-made goal-setting treatments and prevention programmes suited to the specific needs of patients with HMSN1a.

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