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ORIGINAL ARTICLE

Exploratory Study of Physical Activity in Persons With Charcot-Marie-Tooth Disease

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Abstract

Objectives: To explore and describe the perceived facilitators and barriers to physical activity, and to examine the physical activity correlates in people with Charcot-Marie-Tooth (CMT) disease.

Design: Cross-sectional survey study.

Setting: Community-living subjects.

Participants: Swedish people with CMT disease (N=44; men, 54.5%; median age, 59.5y [interquartile range, 45.3–64.8y]).

Interventions: Not applicable.

Main Outcome Measures: The survey included open-ended questions and standardized self-reported scales measuring physical activity, fatigue, activity limitation, self-efficacy for physical activity, fall-related self-efficacy, social support, and enjoyment of physical activity. Physical activity was measured by the Physical Activity Disability Survey–Revised.

Results: Qualitative content analysis revealed that personal factors such as fatigue, poor balance, muscle weakness, and pain were important barriers for physical activity behavior. Facilitators of physical activity were self-efficacy for physical activity, activity-related factors, and assistive devices. Multiple regression analysis showed that self-efficacy for physical activity (β=.41) and fatigue (β=−.30) explained 31.8% of the variation in physical activity ($F_{2,40}=10.78$, $P=.000$).

Conclusions: Despite the well-known benefits of physical activity, physical activity in people with CMT disease is very sparsely studied. These new results contribute to the understanding of factors important for physical activity behavior in people with CMT disease and can guide health professionals to facilitate physical activity behavior in this group of patients.

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Physical activity in people with Charcot-Marie-Tooth (CMT) disease is sparsely studied, despite the well-known benefits of physical activity. The consequences of CMT disease may limit the ability to be physically active. Progressive muscular atrophy of distal muscles, foot deformities, and sensory loss can contribute to altered gait as well as balance problems.[1] As a result, people with the disease frequently trip or fall.[2] In addition, an increased energy cost[3] and fatigue[4] may lead to reduced motivation to be physically active.

Physical activity is defined as all bodily movement that derives from the contraction of the skeletal muscles and results in increased energy expenditure.[5] There is strong evidence that physical activity has positive effects on physical and mental health, quality of life, and the prevention of health problems such as coronary heart disease and type 2 diabetes in the general population.[6-8] In CMT disease, physical training has shown effects on muscle strength, activities of daily living, and balance.[9-13]

Individuals with neuromuscular diseases (including CMT disease) have reported exercise durations shorter than those of control subjects[3,14] and generally represent a sedentary and deconditioned segment of the population.[15]

No prior qualitative study exploring factors important for physical activity behavior in CMT disease have been found. However, in subjects with disabilities, personal motivation, social support, and knowledge of exercise are facilitating factors, whereas barriers include symptoms, fear of injury, fear of falling, and previous negative experiences.[16-20]

Quantitative studies in the general population have shown that health status, self-efficacy for physical activity, age (inversely),

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male sex, educational level, overweight (inversely), and social support are correlates of physical activity.[21] Self-efficacy is defined as the conviction that one can successfully execute the behavior required to produce a desired outcome.[22,23] Both self-efficacy for physical activity and fall-related self-efficacy, defined as the degree of self-efficacy to avoid a fall,[24] are important. Another interesting but sparsely studied factor is enjoyment, a positive affective state that reflects feelings such as pleasure, liking, and fun.[25] Enjoyment is associated with physical activity levels in people with multiple sclerosis (MS).[26] To our knowledge, no previous study has focused on correlates to physical activity in CMT disease.

In persons with CMT disease, as in the general population, physical activity might be one of the most important factors for long-term health and well-being, and more knowledge about physical activity behavior and factors important for this behavior is needed. The aim of this study was to use a qualitative approach to explore and describe the perceived facilitators and barriers to physical activity, and to use a quantitative approach to examine physical activity correlates in persons with CMT disease.

Methods

The study was performed as a cross-sectional cohort survey.

Study cohort

Individuals aged 18 to 81 years with a diagnosis of CMT disease were recruited from neurology clinics in 4 counties in Sweden. All 81 persons registered in the clinics were invited to participate. One person was excluded from answering the survey because of visual problems. The final sample cohort consisted of 44 persons (24 men [54.5%], 20 women), a response rate of 55.0%.

Data collection

After review of the Swedish population register for eligible participants, a questionnaire was sent out by surface mail. An informatory letter, a written consent form, and a stamped reply envelope were included with the questionnaire. One reminder was sent to subjects who did not answer within 3 weeks. All participants in the study provided written informed consent to participate. The study was approved by the Regional Ethical Review Board, Uppsala, Sweden (D-no.: 2010/278).

The questionnaire began with 2 open-ended questions: (1) State the two most important factors that make it easier for you to be physically active; and (2) State the two most important factors that hinder you from being physically active. Thereafter, the Swedish versions of psychometrically sound scales were inserted (described in detail in appendix 1).

The amount of physical activity during the previous week was measured by the Physical Activity Disability Survey—Revised (PADS-R).[27] Activity limitation was measured using the ACTIVLIM questionnaire.[28] Fatigue was measured with the Fatigue Severity Scale.[29,30] Self-efficacy for physical activity was measured using the Exercise Self-Efficacy Scale.[31] Fall-related self-efficacy was measured using the Falls Efficacy Scale.[32] A yes-or-no question on fear of falling was also included (Are you afraid of falling?). Social support for physical activity was measured using Social Influences on Physical Activity.[33] Enjoyment of physical activity was measured using 3 statements on the experience during, or shortly after physical activity of at least 10 minutes' duration (eg, walking). Background variables were also assessed.

List of abbreviations:

CMT	**Charcot-Marie-Tooth**
MS	**multiple sclerosis**
PADS-R	**Physical Activity Disability Survey—Revised**

Data analysis

The written answers from the 2 open-ended questions (qualitative data) were analyzed by inductive content analysis.[34,35] The spontaneous, often very short answers from the subjects were grouped and categorized independently by 2 researchers (K.H., M.E.) according to content similarities. Then the categories were discussed and further developed by the same researchers. The categorization was then reviewed and developed by the first author (E.A.). Finally, the categories and subcategories were discussed and developed by all the authors until consensus was found.

Quantitative data were analyzed using SPSS (version 20[a]). Handling of missing data is shown in appendix 1. Differences between physical activity levels (low physical activity: PADS-R<mean [.056]; high physical activity: PADS-R≥mean [.056]) were analyzed using the chi-square test, Mann-Whitney *U* test or *t* test, depending on the type of scale and distribution, to obtain an understanding of the potential factors affecting physical activity.

Forced-entry multiple regression analysis was used to investigate factors that might influence physical activity (dependent variable). Univariate analyses were first performed with the Pearson correlation coefficient to evaluate the associations between the independent variables and physical activity. Only the 2 independent variables that correlated most to physical activity were included in the multiple regressions analysis because of the low number of participants. No multicollinearity in the regression analysis was found by screening a correlation matrix between all independent variables, and by evaluating the variance inflation factors and tolerance statistics. There was no autocorrelation found with the Durbin-Watson test of independence of residuals. The assumptions of linearity and homoscedasticity were met when visually analyzing the histogram and normal probability plots of the residuals. The level of significance was set at $P<.05$.

Results

Characteristics of total sample

Background characteristics are presented in table 1. The median time since diagnosis was 20 years. In 42% of the participants the self-rated walking distance outdoors was ≤500m. Walking upstairs was impossible or difficult for 31 (70%) of the subjects, and carrying a heavy load was impossible or difficult for 33 (75%) of the subjects. One subject could not walk, 11 (25%) used a walking aid indoors, and 32 (73%) walked indoors without a walking aid.

Of the 36 nonrespondents, 14 (38.9%) were men and 22 were women. No significant difference was found between the sex distributions in the respondents compared with nonrespondents. The median age of nonrespondents was 46.0 years (interquartile range, 35.3—53.8), which was significantly lower ($P=.008$) than the respondents.

Facilitators and barriers to physical activity

Facilitating factors were stated by 34 subjects and barriers by 38 of the subjects (table 2). The 4 emerging categories of the most important perceived facilitators and barriers to physical activity were *activity-related factors, assistive devices, external factors,*