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ORIGINAL ARTICLE

Predictors of participation restriction over a 9-year period in adults with myotonic dystrophy type 1

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ABSTRACT

Purpose: For slowly progressive neuromuscular disease, prognostic approach and long-term monitoring of participation is a crucial part of rehabilitation services. To improve the prognostic approach, professionals must identify individuals at risk of having higher participation restriction. This study aimed to identify personal and environmental predictors of participation restriction over nine years in adults with myotonic dystrophy type 1 (DM1).

Methods: A secondary analysis of a longitudinal design comparing baseline with a follow-up nine years later was used with a multidimensional assessment of participation and personal and environmental factors. Based on theoretical models, multiple linear regressions were used.

Results: One hundred and fourteen adults with DM1 were included in the study (63.2% women; 78.9% adult onset; mean (SD) age of 43.5 (10.4) years). When age, sex, phenotype, and education were controlled for, participation restriction was predicted by a longer time to stand and walk, lower grip strength, higher body mass index, absence of perceived impact of myotonia in daily living, use of adapted transportation from community services, and perception of obstacle in physical environment ($p < 0.001$, adjusted $R^2 = 0.50$).

Conclusions: The majority of predictors of participation restriction can be advantageously modified by rehabilitation and environmental changes, such as politics targeting community services provision or physical environment and services accessibility.

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Health services; longitudinal studies; myotonic dystrophy; patient care management; rehabilitation; social participation

➤ IMPLICATIONS FOR REHABILITATION

- Predictors could better inform rehabilitation professional to recognize individuals at risk of higher participation restriction over time and to target specific interventions based on a prognostic approach.
- Rehabilitation professionals could inform the people living with myotonic dystrophy type 1 and their relatives of the multifactorial nature of occurrence of participation restriction to diminish the "fatality" associated with a genetic progressive disorder.
- Predictors allow professionals to assess and intervene in the management of specific factors depending on the rehabilitation goal.
- Identifying individual with myotonic dystrophy with higher risk of participation restriction could help implement a long-term community based rehabilitation intervention plan targeting both personal and environmental factors.

Introduction

Worldwide, myotonic dystrophy type 1 (DM1) is the most prevalent adult-onset form of muscular dystrophies [1]. DM1 touches 5–13 cases per 100 000 people [2,3] reaching up to 158 per 100 000 in the Saguenay–Lac-Saint-Jean region of northeastern Québec (Canada) due to a founder effect and relative geographical isolation [4,5]. Located on chromosome 19q13.3 [6,7], an excessive repetition in the ADN of cytosine–thymine–guanine (CTG) leads to a progressive cell malfunctioning in multiple organs due to RNA toxicity [8]. Considered as a complex multisystemic disease and often compared to a premature aging process [9–11], DM1 implies heterogeneous symptoms and impairments that greatly varied in terms of presentation and progression [1,12]. While one individual may experience progressive distal to proximal weakness, fatigue, excessive daytime sleepiness, myotonia (i.e., delay in relaxing muscle after voluntary contraction),

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Supplemental data for this article can be accessed here.

pain [13], apathy, depression, and personality disorder [14], another may have, in addition to weakness, the presence of cataracts, cardiac and respiratory impairments, digestive and endocrine deficits [15] and mild cognitive impairments (e.g., attention, visuospatial and constructional disabilities and dysexecutive syndrome) [16]. Adults with DM1 often live in a deprived social environment with lower income, education, and support [17,18], including limited access to social and medical services [19,20]. Due to the dominant inheritance, the social environment of adults with DM1 often implies low social support from family members [17] and a high caregiver burden as he/she often has to look after more than one adult with DM1 [12]. The presence of these personal and environmental factors is likely to influence, even on the long run, several aspects of the life of individuals with DM1 including participation restriction in daily and social activities [21].

Participation restriction is defined by the human development model – disability creation process (HDM-DCP) framework as accomplishment difficulty and/or requirement of assistance in daily and social activities [22]. To guide policy development and service delivery, this model conceptualized participation restriction as a result of a disruptive interaction over time of personal and environmental factors [23]. Apart from the inherent consequences of having difficulty to accomplish daily and social activities, participation restriction lead to direct and indirect consequences in a person's life, such as social isolation, discrimination, stigma, violation of dignity, or financial burden [21]. Associated with quality of life [17,24], participation restriction was reported as a predictor of reduced cognitive functioning [25] and greater risk of mortality [26] in the general aging population. As DM1 is a slowly progressive neuromuscular disease, participation restriction gradually increases over time. A clinically significant increase in restriction has been found in a substantial proportion of adult with DM1 globally (34%), in daily and social activities domains (35% and 38%), and in seven categories: nutrition (34%), fitness (55%), personal care (37%), housing (31%), mobility (44%), community life (36%), and recreation (51%) [27]. To optimize the participation of adults with DM1, medical and rehabilitation long-term follow-up is therefore needed [28], in addition to access to community services and inclusive policies [29].

Long-term monitoring should be a determinant part of rehabilitation services for adults with DM1 [28]. Follow-up and referrals have, however, been often reported as fragmented or happening too late in the process [12,30–32]. In addition, unmet needs for social care and rehabilitation services are reported in a large proportion of adults with DM1 (68–82%) [33] and their relatives (19–31%) [12]. Since 2010, a prognostic approach with anticipatory guidance and health supervision have been advocated to help adults with DM1 in the disease management and optimization of their participation [19]. Annual evaluation of participation in daily and social activities is also recommended [28]. To implement better prognostic approach and to facilitate monitoring of adults with DM1, rehabilitation professionals must identify individuals at risk of having higher participation restriction, such as with long-term predictors. Considering the complex nature of DM1, identifying predictors could help people with their treatment plan and expectations according to disease progression. In addition, environmental factors must be considered as they play a role in the participation restriction (e.g., timely planning of home adaptation to help mobility) and might be more stable landmarks at the society level (e.g., accessibility policies, access to financial assistance, healthcare organization). As environmental factors are considered inseparable from the participation restriction [23], they have the potential to support the long-term management of the disease when identified concurrently with personal predictors. Based on this conceptualization, one hypothesis is that some environmental factors, in addition to personal factors, might act as predictors over a long-term period for participation restriction, especially considering that DM1 is a slowly progressive disease. Considering environmental factors along with personal factors is important to establish a portrait of an individual and identify those at risk of participation restriction over time. They are however sparsely studied in the neuromuscular field, and a knowledge gap to identify their long-term predictors is especially present in DM1. This study thus aimed to identify personal and environmental predictors of participation restriction over nine years in adults with DM1. This paper extends our previous study describing changes in participation over 9-year [27]. Based on the same participant's sample, predictors were specifically identified for the categories of participation that restriction has been found to clinically significantly increase in a substantial proportion of adult with DM1 in our previous study [27].

Methods

Design

A longitudinal study comparing baseline (2002–2004) and follow-up data (2011–2013) was carried out at the Neuromuscular Clinic of the *Centre intégré universitaire de santé et de services sociaux du Saguenay-Lac-Saint-Jean* (Québec, Canada). The current study is a secondary analysis of an interdisciplinary initiative to assess the evolution of multisystemic functions and capabilities, environmental factors, and participation in a large sample of DM1 patients. The study was approved by the Ethics Review Board (Chicoutimi site, #2010-046).

Participants and data collection

At baseline, participants were randomly recruited from the registry of the Neuromuscular Clinic. They were included if they were over 18 years old with a DNA confirmed diagnosis of DM1 with the adult (including juvenile) and late-onset phenotypes. Individuals with congenital or childhood phenotypes or with another condition influencing participation (e.g., stroke) were excluded. A multidimensional clinical assessment inspired by the HDM-DCP was performed by an interdisciplinary team (Figure 1). More specifically, identity factors, motor activity, breathing, and excretion capabilities were assessed during a complete day at the Neuromuscular Clinic by a research assistant, a neurologist, a physiotherapist, and a nurse. In order to minimize fatigue, participation restriction, and environmental factors were assessed at the participant's home during two half-days by an occupational therapist. Finally, a neuropsychologist assessed the intellectual, behavior, and protection and resistance capabilities at the participant's home during two half-days. At follow-up, the baseline' sequence of data collection was kept as similar as possible. French-Canadian version of questionnaires, same examples, and standardized procedures for each test were used at baseline and follow-up.

Variables

Dependent variables: participation restriction

Based on the definition of the HDM-DCP framework, participation restriction was assessed with the short 3.1 version of the Assessment of Life Habits Questionnaire (LIFE-H) [34]. This questionnaire includes 77 activities covering 12 categories of participation (Table 1). Based on self-reported accomplishment level in

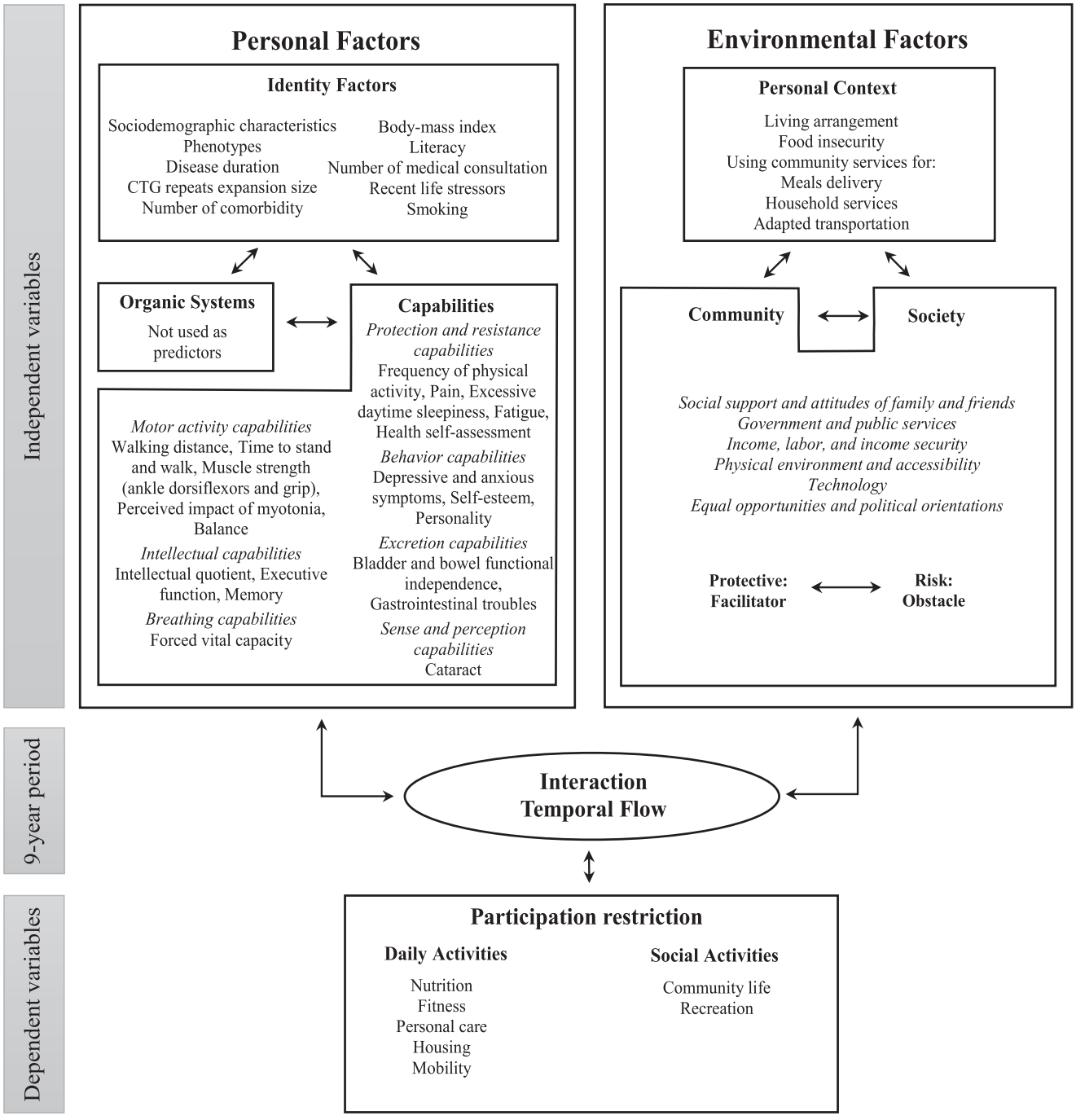

Figure 1. Presentation of the dependent and independent variables collected during the study based on an adapted version of the human development model – disability creation process (HDM-DCP) framework.

activities, which is defined as the difficulty and assistance used to carry out activities, LIFE-H scores range from 0 (not accomplished) to 9 (accomplished without difficulty and assistance). The mention "not applicable" was used when people believed that an activity was non-relevant to them. A mean score is provided for each item, for the total which represents global participation, and for the two domains and each category. The LIFE-H present adequate to excellent psychometric properties for the DM1 population (intra-rater ICC: 0.80–0.91; interrater ICC: 0.86–0.92), except for fitness (intra-rater ICC: 0.20; interrater ICC: 0.21) and communication (intra-rater ICC: 0.12; interrater ICC: 0.47) [35]. Minimal clinically important difference (MCID) was considered with a change of 0.5 (/9) [36].

Identification of independents variables

To identify the independent variables that could potentially predict participation restriction at follow-up (dependent variables), conceptual grounds were first considered [37]. Based on the HDM-DCP framework, theoretical models were built to identify variables best suitable for prediction. An overview of the scientific literature was then carried out by the first author and revised independently by two co-authors, which process is presented in the Supplementary Appendix (Supplementary Material). Thereafter, an interdisciplinary team (psychologist, physiotherapist, biologist, social worker, and research professional specialized in muscular disease) reviewed the theoretical models to add or remove potential predictors to better represent DM1 symptomatology. Theoretical models were

Table 1. Definition and examples of activities for the 12 categories of participation from the HDM-DCP framework [22].

Participation category	Definition	Examples
Daily activities domain		
Nutrition	Activities related to food consumption	Selecting appropriate food for meals; preparing meals; going to restaurants.
Fitness	Activities related to the good shape of the body and mind	Get in and out of bed; sleep; participating in physical activities to maintain fitness.
Personal care	Activities related to a person's bodily well-being	Attending to personal hygiene; dressing; taking care of health.
Communication	Activities related to information exchange with others individual and with collectivity	Talking; writing; using a phone.
Housing	Activities related to a person's residence	Choosing and maintaining a home; doing household tasks; entering and exiting home; moving outside around home.
Mobility	Activities related to trips over short or long distances with or without means of transport	Getting around on slippery or uneven surfaces; driving a vehicle; riding a bicycle.
Social activities domain		
Responsibilities	Activities related to the assumption of financial, civil, and family responsibilities	Using money; elaborating a budget; Go vote; taking care of children or family member.
Interpersonal relationship	Activities related to relationships with others	Maintaining a close relationship with partner; maintaining friendships; having a sexual relationship.
Community life	Activities related to consumption of goods and services of community	Getting to, entering, and using public buildings in community; taking part in associative life; participating in spiritual or religious practices.
Education	Activities related to psychomotor, intellectual, social, and cultural development	Attending to school; receiving general training.
Employment	Activities related to a person's principal occupation	Working; volunteering.
Recreation	Activities related to recreational, cultural or sporting activities during free time in a context of fun and liberty	Going to artistic or cultural events; participating in tourist activities; taking part in outdoor activities.

developed according to the definitions and seven categories of participation of the HDM-DCP framework (Supplementary Table S1). The theoretical models led to the identification of overall 56 potential independent variables among the personal and environmental factors (Figure 1). When available, objective measures were privileged. Among the independent variables, five were identified as potential confounders based on previous work [38–41]. The team decided, in addition to test CTG repeats expansion size as a predictor, to see it potential to explain participation restriction over time.

Independent variables: personal and environmental factors

Personal factors included identity factors, motor activity, intellectual, breathing, protection and resistance, behavior, excretion, and sense and perception capabilities (Figure 1). Environmental factors included personal context and community and society factors (Figure 1). The definition of variables and psychometric proprieties of measurement tools are presented in the Supplemental Appendix (Supplementary Material). Measurement tools presented adequate to excellent psychometric proprieties (references provided in Supplementary Material), mostly for neuromuscular or DM1 adults.

Data analysis

Participant characteristics were shown with mean (SD) for continuous variables and frequency (%) for categorical variables. To assess change in patients' characteristics between T1 and T2, paired *t* tests, Wilcoxon's signed ranks, and McNemar's tests were used. Among the 56, eight independent variables presented missing data of less than 15% that were accommodated at the composite score [42] with five multiple imputations as recommended by Graham et al. [43]. While the majority ($n = 81$; 71.1%) of respondents had no missing data at baseline, less than a quarter ($n = 27$; 23.7%) presented one or two and few others ($n = 6$; 5.3%) had at most five missing values. As data were extracted from the medical chart rather than collected for this study or perform on a subset for time resource consideration, disease duration, BBS, and NEO-FFI presented more than 15% missing values. These three variables were excluded to preserve the stability of the modeling [44] and avoid the decrease of power under 80% with the use of multiple imputations [43]. Multivariate linear regression was used to predict participation restriction after a 9-year period with independent variables at baseline. As the focus was made on restriction, participation scores were inverted (–9 to 0) to facilitate the interpretation of the regression model. Regression models were built with a three-step process. First, based on theoretical models, univariate linear regression was performed to identify the best predictors among independent variables for each category of participation (Supplementary Table S1). Univariate models for global participation as well as daily and social activities domains were built based on significant independent variables extracted from the composing category of participation (data not shown). All variables with alpha $\geq$0.10 were excluded from the subsequent analysis regarding the exploratory process [45]. Second, multivariate regression models were built for personal and environmental factors separately. Regardless of their level of significance, confounding variables (i.e., age, sex, phenotype, and education at baseline) were first forced into the models considering their theoretical importance. Due to the high number of potential univariate predictors, a hierarchical method based on clinical reasoning was used to select important variables to put in the multivariate model with stepwise strategy (Supplementary Tables S2–S11). Hierarchical method consisted of selecting first the potential personal and environmental factors with alpha $<$0.10 that correlated more strongly with participation restriction in each subcategory (e.g., higher fatigue before bad health self-assessment). Third, final models were built with significant independent variables from each personal and environmental factors with a chunkwise strategy (first chunk: confounding variables, second chunk: personal factors, and third chunk: environmental factors). Most parsimonious final models were identified considering the best adjusted R^2, standardized coefficient, and lower confidence intervals. As they were found to be potentially

modifying factors [27,38,46], interaction terms of sex with grip strength was tested in final models when applicable. Linearity, homoscedasticity, and normality assumptions were tested on final models with a graphical review of standardized predicted values on studentized residuals as well as with Kolmogorov–Smirnov's analysis. Multicollinearity was also tested with the variance inflation factor. Post hoc power analysis was performed with NQuery Advisor 6 software prior to analysis. Based on an alpha significance level of 5% and a power of 80%, a sample size of 95 people allowed detection of an R^2 of 0.2000 in a multiple linear regression test of $R^2 = 0$ for 11 variables. With a sample size of 114, multiple linear regression analysis will have 96% power to detect a medium to large effect size ($f^2 = 0.2$). Considering the hierarchical integration, a maximum of 11 variables was considered at the same time in each intermediary model. With 11 variables, the ratio variable/observation was 10, which is frequently deemed acceptable [45]. The statistical analyses were performed using SPSS software (version 25.0 for Windows; SPSS Inc., Chicago, IL).

Results

Characteristics of participants

A total of 115 participants was included in this follow-up study (Figure 2), but one man was excluded during the analysis because he presented atypical scores (i.e., outliers) likely caused by highly

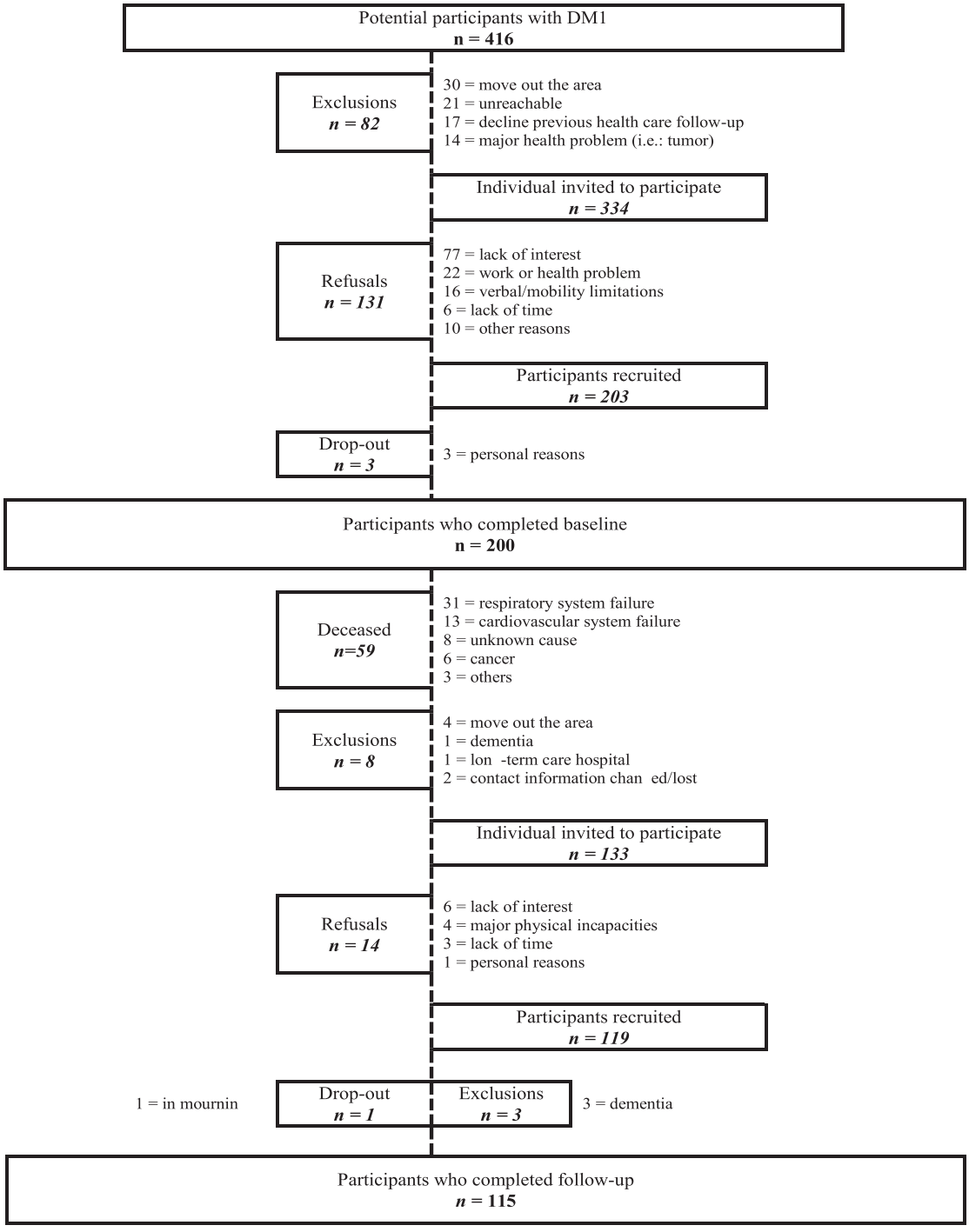

Figure 2. Participants' flowchart. Already published in Raymond et al. [27] (reuse permission granted).

Table 2. Characteristics of DM1 participants ($n = 114$).

Personal factors	Baseline	Follow-up	*p* Value
Identity factors			
Age (years)	43.5 (10.4)	52.2 (10.3)	<0.001
Sex (women = 0)	72 (63.2)		
Education (years)			
≤11	50 (43.9)	50 (43.9)	0.91
12–13	48 (42.1)	48 (42.1)	
14–16	14 (12.3)	12 (10.5)	
≥17	2 (1.7)	4 (3.5)	
Family income (Canadian $)			
<10 000	20 (17.5)	11 (9.7)	0.28
10 000–19 999	38 (33.3)	51 (44.7)	
20 000–39 999	19 (16.7)	23 (20.2)	
40 000–59 999	10 (8.8)	16 (14.0)	
>60 000	18 (15.8)	12 (10.5)	
Unknown/refused	9 (7.9)	1 (0.9)	
Marital status			
Married	43 (37.7)	55 (48.2)	<0.001
Divorced or widowed	12 (10.5)	21 (18.4)	
Single	59 (51.8)	38 (33.3)	
Phenotype (adult/juvenile = 0)	90 (78.9)		
Disease duration [min–max] Ø	19.9 (8.1) [3–38]		
Missing data, *n* (%)	33 (28.9)		
CTG repeats expansion size	777 (516)	923 (505)	<0.001
Missing data, *n* (%)	–	2 (1.8)	
Number of comorbidity	1.8 (1.6)	–	
Body mass index	25.1 (5.4)	25.7 (5.9)	0.03
Literacy for filing form			
No help needed	93 (81.6)	87 (76.3)	<0.01
Needed help sometimes	14 (12.3)	6 (5.3)	
Needed help often	7 (6.1)	21 (18.4)	
Number of medical consultation during last year	4.0 (18.1)	2.3 (5.1)	0.32
Recent life stressors (no = 0)	101 (88.6)	–	
Active smoking (yes = 1)	37 (32.4)	28 (24.6)	<0.001
Motor activity capabilities			
Walking distance (2MWT, m)	135.2 (27.2)	–	
Time to stand and walk (TUG, s)	10.1 (2.0)	10.1 (3.9)	0.24
Missing data, *n* (%)	–	16 (14.0)	
Ankle dorsiflexors strength (QMT, Nm)	16.4 (7.8)	7.4 (4.8)	<0.001
Missing data, *n* (%)	8 (7.0)	7 (6.1)	
Dominant grip strength (JAMAR, kg)	12.2 (10.1)	10.9 (9.2)	<0.001
Missing data, *n* (%)	1 (0.9)	4 (3.5)	
Perceived impact of myotonia (yes = 1)	46 (40.4)	–	
Balance (BBS, /56) Ø	52.9 (6.8)	47.1 (13.6)	<0.001
Missing data, *n* (%)	30 (26.3)	7 (6.1)	
Intellectual capabilities			
Intellectual quotient (WAIS-R, full scale)	83.1 (8.4)	84.9 (9.0)	<0.001
Missing data, *n* (%)	3 (2.6)	2 (1.8)	
Executive function (SCWT, *T*-score)	46.9 (6.9)	49.9 (7.5)	<0.001
Missing data, *n* (%)	5 (4.4)	4 (3.5)	
Memory (CVLT, *Z*-score)	0.19 (1.1)	–0.5 (1.2)	<0.001
Missing data, *n* (%)	–	1 (0.9)	
Breathing capabilities			
Forced vital capacity (spirometer, in L)	3.1 (0.9)	2.7 (0.9)	<0.001
Missing data, *n* (%)	15 (13.2)	15 (13.2)	
Protection and resistance capabilities			
Frequency of physical activity			
Never	39 (34.2)	62 (54.4)	0.04
1 time a month	11 (9.6)	3 (2.6)	
2–3 times a month	7 (6.1)	8 (7.0)	
1 time a week	14 (12.3)	6 (5.3)	
2 times a week	10 (8.8)	6 (5.3)	
3 times a week	18 (15.8)	5 (4.4)	
4 times and more a week	15 (13.2)	24 (21.1)	
Pain (yes = 1)	83 (72.8)	–	
Daytime sleepiness (DSS, /15)	4.5 (2.9)	5.3 (3.5)	<0.01
Missing data, *n* (%)	–	2 (1.8)	
Fatigue (KFSS, /63)	39.1 (15.3)	43.4 (15.4)	<0.001
Missing data, *n* (%)	–	1 (0.9)	
Actual health self-assessment			
Bad to passable	30 (26.3)	44 (38.6)	<0.001
Good	43 (37.7)	45 (39.5)	
Excellent to very excellent	41 (36.0)	25 (21.9)	
Behavior capabilities			
Depression symptoms (SCL-90, *T*-score)	50.8 (9.1)	51.5 (9.0)	0.41
Missing data, *n* (%)	11 (9.6)	1 (0.9)	

(*continued*)

Table 2. Continued.

Personal factors	Baseline	Follow-up	*p* Value
Anxiety symptoms (SCL-90, *T*-score)	46.6 (9.3)	46.2 (8.8)	0.96
Missing data, *n* (%)	10 (8.8)	1 (0.9)	
Self-esteem (Rosenberg, /40)	31.0 (4.5)	30.2 (5.2)	0.07
Missing data, *n* (%)	1 (0.9)	–	
Personality traits (NEO-FFI, *T*-score) Ø			
Neuroticism	48.2 (9.3)	50.6 (10.2)	0.20
Extraversion	50.0 (8.6)	47.8 (9.8)	0.01
Openness	41.3 (7.5)	41.3 (8.2)	0.52
Agreeableness	49.2 (9.4)	48.0 (9.0)	0.17
Conscientiousness	50.3 (8.7)	46.5 (8.1)	<0.001
Missing data, *n* (%)	43 (37.7)	1 (0.9)	
Excretion capabilities			
Functional independence (FIM, /7)			
Bladder management	6.6 (0.9)	–	
Bowel management	6.7 (0.8)	–	
Gastrointestinal disturbances (yes = 1)	62 (54.4)	–	
Sense and perception capabilities			
Cataract (yes = 1)	47 (41.2)	32 (28.1)	<0.001
Environmental factors			
Personal context			
Living arrangement (home alone = 0)	18 (15.8)	33 (28.9)	<0.01
Food insecurity (no)	96 (84.2)	99 (86.8)	0.65
Floor of living area (ground level = 0)	37 (32.5)	71 (62.3)	<0.001
Use of community services (yes = 1)			
Meals delivery	15 (13.2)	8 (7.0)	0.27
Household assistance	50 (43.9)	56 (49.1)	0.29
Adapted transportation	21 (18.4)	26 (22.8)	0.54
Community and society (MQE)			
Social support and attitude			
Obstacle	0.5 (0.9)	–	
Facilitator	10.5 (6.3)	–	
Income, labor, and income security			
Obstacle	1.6 (2.1)	–	
Facilitator	8.1 (4.2)	–	
Government and public services			
Obstacle	1.0 (1.6)	–	
Facilitator	12.7 (5.2)	–	
Physical environment and accessibility			
Obstacle	14.1 (9.5)	–	
Facilitator	8.0 (7.1)	–	
Technology			
Obstacle	0.4 (1.0)	–	
Facilitator	2.5 (2.1)	–	
Equal opportunities and political orientations			
Obstacle	1.8 (2.6)	–	
Facilitator	1.2 (1.5)	–	
Participation restriction (LIFE-H, /-9)			
Global participation	−8.2 (0.8)	−7.7 (1.2)[b]	<0.001
Daily activities	−8.2 (0.8)	−7.8 (1.2)	<0.001
Nutrition ($n = 112$)[a]	−8.6 (0.8)	−7.8 (1.8)[b]	<0.001
Fitness	−8.1 (1.2)	−7.1 (1.7)[b]	<0.001
Personal care	−8.7 (0.5)	−8.0 (1.4)[b]	<0.001
Housing	−7.5 (1.5)	−7.4 (1.5)	0.50
Mobility	−7.5 (1.7)	−7.0 (2.1)[b]	<0.01
Social activities	−8.2 (0.9)	−7.7 (1.4)[b]	<0.001
Community life	−8.5 (1.0)	−7.7 (2.1)[b]	<0.001
Recreation ($n = 111$)[a]	−7.3 (2.5)	−5.8 (3.0)[b]	<0.001

Abbreviation. (precision on scoring interpretation): 2MWT: 2-minute walk test (a higher score indicated a longer walking distance); BBS: Berg Balance Scale (a higher score indicated a higher balance); BMI: body mass index; CTG: cytosine–thymine–guanine; CVLT: California Verbal Learning Test (*Z*-score of 0 ± 1 indicated an average memory); DM1: myotonic dystrophy type 1; DSS: Daytime Sleepiness Scale (score of ≥7 indicated excessive daytime sleepiness); FIM: Functional Independence Measure (higher score indicated higher functional independence); JAMAR: Jamar dynamometer (a higher score indicated a higher strength); KFSS: Krupp Fatigue Severity Scale (score of ≥36 indicated a greater fatigue); LIFE-H: Assessment of Life Habits Questionnaire (inversed score of −9 to 0, with −9 indicating less participation restriction); MQE: Measure of the Quality of the Environment (obstacle scoring were positively reported, a higher score indicated perception of major obstacle or facilitator); NEO-FFI: NEO Five-Factor Inventory (*T*-score of 50 ± 10 indicated an average personality trait); QMT: quantitative muscular testing (a higher score indicated a higher strength); RSES: Rosenberg Self-esteem Scale (higher score indicated higher self-esteem); SCL-90: Symptom Checklist-90 Revised (*T*-score of 50 ± 10 indicated an average symptomatology); SCWT: Stroop Color and Word Test (*T*-score of 50 ± 10 indicated an average executive function); TUG: Timed-up and go (a higher score indicated a lower mobility); WAIS-R: Wechsler Adult Intelligence Scale-Revised (a mean score of 100 ± 15 is considered normal intellectual quotient).

Values expressed as mean (SD) for continuous variables and frequency (%) for categorical variables.

Ø refers to variables not included in the regression models.

[a]Due to the possibility to score "not applicable" activity in the LIFE-H, nutrition and recreation presented lower sample.

[b]Change of 0.5 points is clinically significant.

Table 3. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of *global participation* restriction ($N = 114$).

	Model 1 (adjusted R^2=0.19)		Model 2 (adjusted R^2=0.43)		Model 3 (adjusted R^2=0.50)		Model without confounding (adjusted R^2=0.47)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−8.39 ± 0.74	<0.001	−9.85 ± 0.85	<0.001	−10.47 ± 0.82	<0.001	−10.29 ± 0.63	<0.001
Age	0.03 ± 0.01	<0.01	0.01 ± 0.01	0.57	0.01 ± 0.01	0.35		
Sex	0.26 ± 0.22	0.23	0.52 ± 0.19	<0.01	0.49 ± 0.18	<0.01		
Phenotype	−1.32 ± 0.29	<0.001	−0.05 ± 0.35	0.88	−0.09 ± 0.32	0.79		
Education	−0.06 ± 0.04	0.16	−0.01 ± 0.04	0.74	−0.01 ± 0.03	0.75		
Body mass index		0.05 ± 0.02	<0.01	0.03 ± 0.02	0.04	0.04 ± 0.02	0.02	
Time to stand and walk			0.15 ± 0.05	<0.01	0.13 ± 0.05	<0.01	0.14 ± 0.05	<0.01
Grip strength			−0.05 ± 0.01	<0.001	−0.04 ± 0.01	<0.01	−0.03 ± 0.01	<0.001
Perceived impact of myotonia			−0.56 ± 0.19	<0.01	−0.47 ± 0.18	0.01	−0.51 ± 0.18	<0.01
Use of community services of adapted transportation					0.41 ± 0.13	<0.01	0.44 ± 0.13	0.001
Perception of physical environment and accessibility as obstacle					0.02 ± 0.01	0.03	0.02 ± 0.01	0.06

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

Table 4. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *daily activities* ($N = 114$).

	Model 1 (adjusted R^2=0.22)		Model 2 (adjusted R^2=0.48)		Model 3 (adjusted R^2=0.54)		Model without confounding (adjusted R^2=0.50)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−8.39 ± 0.70	<0.001	−10.15 ± 0.79	<0.001	−10.72 ± 0.76	<0.001	−10.68 ± 0.59	<0.001
Age	0.04 ± 0.01	0.001	0.01 ± 0.01	0.49	0.01 ± 0.01	0.30		
Sex	0.33 ± 0.21	0.11	0.59 ± 0.18	0.001	0.56 ± 0.17	0.001		
Phenotype	−1.27 ± 0.28	<0.001	0.05 ± 0.32	0.88	0.02 ± 0.30	0.95		
Education	−0.07 ± 0.04	0.07	−0.03 ± 0.03	0.46	−0.02 ± 0.03	0.46		
Body mass index		0.04 ± 0.02	<0.01	0.03 ± 0.01	0.04	0.04 ± 0.02	0.02	
Time to stand and walk		0.18 ± 0.05	<0.001	0.16 ± 0.04	<0.001	0.18 ± 0.04	<0.001	
Grip strength			−0.05 ± 0.01	<0.001	−0.04 ± 0.01	0.001	−0.03 ± 0.01	0.001
Perceived impact of myotonia		−0.43 ± 0.17	0.01	−0.35 ± 0.17	0.04	−0.42 ± 0.17	0.02	
Use of community services of adapted transportation		0.42 ± 0.12	0.001	0.44 ± 0.13	<0.001			
Perception of physical environment and accessibility as obstacle		0.02 ± 0.01	0.048	0.02 ± 0.01	0.11			

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

uncontrolled diabetes. Compared to the 114 who participated in follow-up, the 85 adults who did not participate in follow-up did not differ for sex, CTG repeats, and phenotypic distribution. Yet, they were older (51.6 ± 12.1 y.o., $p < 0.01$) and had more participation restriction (−7.3 ± 1.2, $p < 0.01$). Aged between 20 and 77 years at baseline, participants were mostly women and with adult phenotype (Table 2). Most participants were living at home with a spouse or other relatives and had a family income of less than C$20k. At baseline, all participants were able to walk for short or long distances (71.1–230.6 m during 2 min), only four were using technical aids (can or roller walker, data not shown). The mean score of the participants' group falls slightly below average intellectual quotient. However, executive and memory functioning were in the normal range (Table 2). The environment was perceived mostly as a facilitator for social support and attitude of family and friends, income, labor and income security, government and public services, and physical environment and accessibility. The latter was also perceived as a major obstacle. Among the half of the participants (49.1%) that moved during the study, two-thirds (60.7%) moved at ground level. The others moved in a similar living area. Globally, participants accomplished their daily and social activities without difficulty but using assistive devices or adaptation. Participation restriction increased clinically significantly over time for all categories, except housing and daily activities domain. Restricted categories were in decreasing order: recreation, mobility, fitness, housing, community life, social activities domain, global participation, nutrition, daily activities domain, and personal care.

Best predictors of participation restriction over nine years

Global participation

When controlling for potential confounding variables, half of the variance of global participation was explained (Table 3). In fact, a higher BMI, a longer time to stand and walk, a lower grip strength, not perceiving impact of myotonia, use of community services of adapted transportation, and perception of physical environment as obstacle predicted greater global participation restriction over time (Table 3). Without controlling for confounding variables, predictors explain 47% of the variance. When removing the confounding variables, little meaningfully changes

Table 5. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *nutrition* ($N = 112$).[a]

	Model 1 (adjusted R^2=0.14)		Model 2 (adjusted R^2=0.42)		Model without confounding (adjusted R^2=0.35)	
	β±SE	*p* Value	β±SE	*p* Value	β±SE	*p* Value
Intercept	−8.49 ± 1.13	<0.001	−8.38 ± 1.73	<0.001	−8.86 ± 1.43	<0.001
Age	0.05 ± 0.02	<0.01	0.03 ± 0.02	0.09		
Sex	0.24 ± 0.33	0.46	0.38 ± 0.29	0.19		
Phenotype	−1.20 ± 0.46	<0.01	0.57 ± 0.46	0.22		
Education	−0.14 ± 0.07	0.03	−0.09 ± 0.06	0.09		
CTG repeats expansion size		0.001 ± 0.0003	<0.001	0.001 ± 0.0003	<0.01	
Time to stand and walk		0.26 ± 0.08	<0.01	0.33 ± 0.07	<0.001	
Functional independence for bowel management	−0.53 ± 0.17	<0.01	−0.45 ± 0.18	0.01		

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.
[a]Two participants indicated all nutrition activities as "not applicable" to them.

Table 6. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *fitness* ($N = 114$).

	Model 1 (adjusted R^2=0.13)		Model 2 (adjusted R^2=0.30)		Model without confounding (adjusted R^2=0.14)	
	β±SE	*p* Value	β±SE	*p* Value	β±SE	*p* Value
Intercept	−11.76 ± 1.64	<0.001	−10.30 ± 1.95	<0.001	−7.78 ± 0.70	<0.001
Age	0.02 ± 0.02	0.22	0.01 ± 0.02	0.59		
Sex	0.57 ± 0.32	0.07	1.36 ± 0.38	<0.001		
Phenotype	1.65 ± 0.43	<0.001	0.72 ± 0.47	0.13		
Education	0.06 ± 0.06	0.37	0.11 ± 0.06	0.05		
Forced vital capacity			−0.58 ± 0.24	0.02	−0.25 ± 0.18	0.17
Fatigue			0.04 ± 0.01	<0.001	0.04 ± 0.01	<0.001

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

Table 7. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *personal care* ($N = 114$).

	Model 1 (adjusted R^2=0.14)		Model 2 (adjusted R^2=0.41)		Model 3 (adjusted R^2=0.49)		Model without confounding (adjusted R^2=0.45)	
	β±SE	*p* Value	β±SE	*p* Value	β±SE	*p* Value	β±SE	*p* Value
Intercept	−8.52 ± 0.89	<0.001	−11.07 ± 0.92	<0.001	−10.46 ± 0.88	<0.001	−10.95 ± 0.62	<0.001
Age	0.04 ± 0.01	<0.01	−0.002 ± 0.01	0.84	−0.004 ± 0.01	0.69		
Sex	0.37 ± 0.26	0.16	0.70 ± 0.23	0.002	0.76 ± 0.21	<0.001		
Phenotype	−1.22 ± 0.36	0.001	0.35 ± 0.41	0.39	0.48 ± 0.38	0.21		
Education	−0.09 ± 0.05	0.09	−0.04 ± 0.04	0.35	−0.04 ± 0.04	0.29		
Time to stand and walk			0.35 ± 0.06	<0.001	0.23 ± 0.06	<0.001	0.24 ± 0.06	<0.001
Grip strength			−0.04 ± 0.01	<0.01	−0.03 ± 0.01	0.04	−0.01 ± 0.01	0.20
Perception of physical environment and accessibility as facilitator				0.05 ± 0.02	< 0.01	0.05 ± 0.02	<0.01	
Perception of technology as facilitator					0.13 ± 0.05	0.01	0.14 ± 0.05	0.01

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

in coefficient estimates were observed for a few variables (e.g., perceived impact of myotonia: −0.47 to −0.51).

Daily activities

After controlling for confounding variables, predictors of participation restriction in daily activities domain explained 54% of the variance of the model at follow-up (Table 4). A higher BMI, a longer time to stand and walk, a lower grip strength, not perceiving impact of myotonia, use of community services of adapted transportation, and perception of physical environment as obstacle predicted greater participation restriction over time in daily activities (Table 4).

For all daily activities' category, each model explained between 30% and 49% of the variance of participation restriction at follow-up (Tables 5–9).

Nutrition and fitness. When confounding variables were controlled for, a higher CTG repeats expansion size, a longer time to stand and walk, and a lower functional independence for bowel management predicted greater participation restriction in nutrition (Table 5). However, a lower forced vital capacity and a higher perceived fatigue predicted greater participation restriction in fitness (Table 6). Although environmental factors were considered (Supplementary Tables S3 and S4), only personal factors significantly contributed to predicting participation restriction in nutrition and fitness over time.

Personal care. After controlling for confounding variables, a longer time to stand and walk, a lower grip strength, and perception of physical environment and technology as facilitators predicted greater participation restriction in personal care (Table 7).

Housing. After controlling for confounding variables, a shorter walking distance and a higher perceived fatigue, not living at ground level, and perception of physical environment as obstacle predicted greater participation restriction in housing (Table 8).

Mobility. After controlling for confounding variables, a higher BMI, a lower grip strength, not perceiving impact of myotonia, and use of community services of adapted transportation predicted greater participation restriction in mobility (Table 9).

Table 8. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *housing* ($N = 114$).

	Model 1 (adjusted R^2=0.19)		Model 2 (adjusted R^2=0.38)		Model 3 (adjusted R^2=0.43)		Model without confounding (adjusted R^2=0.37)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−8.22 ± 0.92	<0.001	−6.64 ± 1.27	<0.001	−7.59 ± 1.28	<0.001	−6.02 ± 0.80	<0.001
Age	0.04 ± 0.01	<0.01	0.03 ± 0.01	0.03	0.03 ± 0.01	0.01		
Sex	0.08 ± 0.27	0.77	0.40 ± 0.24	0.09	0.38 ± 0.23	0.10		
Phenotype	−1.74 ± 0.37	<0.001	−0.91 ± 0.35	0.01	−0.78 ± 0.35	0.02		
Education	−0.07 ± 0.05	0.19	−0.05 ± 0.05	0.31	−0.05 ± 0.05	0.28		
Walking distance			−0.02 ± 0.003	<0.001	−0.02 ± 0.003	<0.001	−0.02 ± 0.004	<0.001
Fatigue			0.03 ± 0.01	0.001	0.02 ± 0.01	0.03	0.02 ± 0.01	0.08
Not living at ground level					0.55 ± 0.23	0.02	0.48 ± 0.24	0.044
Perception of physical environment and accessibility as obstacle				0.03 ± 0.01	0.02	0.04 ± 0.01	<0.01	

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

Table 9. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *mobility* ($N = 114$).

	Model 1 (adjusted R^2=0.17)		Model 2 (adjusted R^2=0.31)		Model 3 (adjusted R^2=0.34)		Model without confounding (adjusted R^2=0.34)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−7.21 ± 1.28	<0.001	−8.04 ± 1.41	<0.001	−8.60 ± 1.40	<0.001	−8.47 ± 0.82	<0.001
Age	0.04 ± 0.02	0.03	0.02 ± 0.02	0.39	0.02 ± 0.02	0.39		
Sex	−0.05 ± 0.38	0.89	0.22 ± 0.36	0.53	0.18 ± 0.35	0.61		
Phenotype	−2.33 ± 0.51	<0.001	−0.90 ± 0.64	0.16	−0.89 ± 0.63	0.16		
Education	−0.11 ± 0.07	0.15	−0.05 ± 0.07	0.48	−0.04 ± 0.07	0.54		
Body mass index			0.09 ± 0.03	<0.01	0.08 ± 0.03	<0.01	0.09 ± 0.03	<0.01
Grip strength			−0.07 ± 0.02	0.001	−0.07 ± 0.02	<0.01	−0.09 ± 0.02	<0.001
Perceived impact of myotonia			−0.99 ± 0.35	0.004	−1.01 ± 0.34	<0.01	−1.04 ± 0.33	0.001
Use of community services of adapted transportation				0.57 ± 0.26	0.03	0.60 ± 0.26	0.02	

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

Social activities

After controlling for confounding variables, predictors of participation restriction in social activities domain explained 39% of the variance of the model at follow-up (Table 10). A lower family income and grip strength, and perception of physical environment as obstacle predicted greater participation restriction over time in social activities domain (Table 10). In addition, sex interacted with grip strength by modifying the effect of sex in the model which the coefficient changes from positive and statistically significant to negative and not significant.

For community life and recreation category, model explained respectively 20% and 52% of the variance of participation restriction at follow-up (Tables 11 and 12).

Community life. A shorter walking distance and perception of physical environment as obstacle predicted greater participation restriction in community life (Table 11). However, community life was the category with the lowest level of explained variance.

Recreation. A lower family income and grip strength, not perceiving impact of myotonia, a lower memory, and perception of equal opportunities and political orientations as obstacle predicted greater participation restriction in recreation (Table 12). In addition, sex interacted with grip strength by modifying the effect of sex the same way did the social activities domain.

Many predictors were recurrent from one participation category to another (Table 13). However, the most recurrent predictors of participation restriction were in order of importance: for *personal* factors, lower grip strength, longer time to stand and walk, perceiving impact of myotonia in daily living, and higher BMI; and, for *environmental* factors, perception of obstacle in physical environment and accessibility and use of community services of adapted transportation.

Discussion

This study identified personal and environmental predictors of participation restriction over a 9-year period. To our knowledge, this is the first study to identify predictors of long-term participation based on a holistic conceptual framework for adults with DM1. The cohort was similar in term of age at baseline from the adult phenotype of the DM-Scope registry in Europe (46.7 ± 10.0 y.o.), but slightly differ in terms of sex proportion (55% women) and CTG (±500 repeats) [15]. Predictors slightly differ between global participation, daily activities domain, and social activities domains. Although knowledge of both personal and environmental predictors are crucial to implement a prognostic approach [19], very few studies addressed prediction of participation over a longitudinal course in slowly progressive neuromuscular disorders. Similarly to the present results, Kalkman et al. found that strength weakness, less physical activity, sleep disturbance, pain, and higher fatigue were predictors of greater global activity limitation over 18 months for 198 participants with three neuromuscular disorders, including DM1 [47]. Psychological distress, neuropsychological impairment, social functioning along with social support were not retained in Kalkman's model. However, they did not use theoretical models to guide their analysis and, as their principal focus was to predict fatigue and activity limitations, they did not included much environmental predictors, except social support. Differences could also be partially explained by the shorter time of Kalkman's study and a different operationalization of the participation. To our knowledge,

Table 10. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *social activities* ($N = 114$).

	Model 1 (adjusted R^2=0.15)		Model 2 (adjusted R^2=0.31)		Model 3 (adjusted R^2=0.36)		Model 4 (adjusted R^2=0.39)		Model without confounding (adjusted R^2=0.37)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−8.52 ± 0.87	<0.001	−7.64 ± 0.80	<0.001	−8.45 ± 0.82	<0.001	−7.57 ± 0.88	<0.001	−7.09 ± 0.35	<0.001
Age	0.04 ± 0.01	0.01	0.03 ± 0.01	0.02	0.03 ± 0.01	0.02	0.02 ± 0.01	0.12		
Sex	0.24 ± 0.26	0.36	0.51 ± 0.25	0.04	0.50 ± 0.24	0.04	−0.15 ± 0.35	0.67		
Phenotype	−1.33 ± 0.35	<0.001	0.16 ± 0.44	0.71	0.03 ± 0.43	0.95	0.56 ± 0.47	0.23		
Education	−0.05 ± 0.05	0.33	−0.01 ± 0.05	0.76	−0.01 ± 0.04	0.83	−0.001 ± 0.04	0.97		
Family income			−0.14 ± 0.05	<0.01	−0.12 ± 0.05	<0.01	−0.12 ± 0.04	<0.01	−0.11 ± 0.04	0.01
Grip strength			−0.06 ± 0.02	<0.001	−0.04 ± 0.02	0.02	−0.10 ± 0.03	0.01	−0.07 ± 0.02	<0.001
Perception of physical environment and accessibility as obstacle			0.04 ± 0.01	<0.01	0.03 ± 0.01	0.01	0.04 ± 0.01	<0.01		
Sex × grip strength							0.06 ± 0.02	0.02	0.05 ± 0.01	0.001

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

Table 11. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *community life* ($N = 114$).

	Model 1 (adjusted R^2=0.09)		Model 2 (adjusted R^2=0.14)		Model 3 (adjusted R^2=0.20)		Model without confounding (adjusted R^2=0.18)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−8.78 ± 1.33	<0.001	−5.45 ± 1.77	<0.01	−6.91 ± 1.79	<0.001	−5.62 ± 1.02	<0.001
Age	0.05 ± 0.02	0.02	0.03 ± 0.02	0.15	0.03 ± 0.02	0.13		
Sex	0.11 ± 0.39	0.78	0.33 ± 0.39	0.40	0.42 ± 0.38	0.26		
Phenotype	−1.80 ± 0.53	0.001	−1.12 ± 0.57	0.049	−0.80 ± 0.56	0.16		
Education	−0.06 ± 0.08	0.43	−0.06 ± 0.07	0.40	−0.05 ± 0.07	0.48		
Walking distance			−0.02 ± 0.01	<0.01	−0.02 ± 0.01	0.02	−0.02 ± 0.01	0.001
Perception of physical environment and accessibility as obstacle			0.06 ± 0.02	<0.01	0.06 ± 0.02	<0.01		

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.

Table 12. Unstandardized regression coefficients in the final multivariate model identifying with chunkwise strategy the best predictors of participation restriction in *recreation* ($N = 111$)[a].

	Model 1 (adjusted R^2=0.18)		Model 2 (adjusted R^2=0.44)		Model 3 (adjusted R^2=0.49)		Model 4 (adjusted R^2=0.52)		Model without confounding (adjusted R^2=0.52)	
	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value	β±SE	p Value
Intercept	−6.96 ± 1.88	0.001	−4.68 ± 1.64	<0.01	−5.20 ± 1.57	<0.01	−3.86 ± 1.63	0.02	−2.34 ± 0.50	<0.001
Age	0.06 ± 0.03	0.06	0.03 ± 0.03	0.25	0.03 ± 0.02	0.18	0.02 ± 0.02	0.48		
Sex	0.42 ± 0.56	0.46	1.02 ± 0.49	0.04	0.98 ± 0.47	0.04	−0.27 ± 0.70	0.70		
Phenotype	−3.39 ± 0.75	0.001	−0.60 ± 0.87	0.49	−0.87 ± 0.84	0.30	0.13 ± 0.92	0.88		
Education	−0.07 ± 0.11	0.54	0.10 ± 0.10	0.28	0.08 ± 0.09	0.37	0.10 ± 0.09	0.26		
Family income			−0.30 ± 0.09	0.001	−0.29 ± 0.09	<0.01	−0.29 ± 0.09	0.001	−0.27 ± 0.08	<0.01
Grip strength			−0.12 ± 0.03	<0.001	−0.12 ± 0.03	<0.001	−0.22 ± 0.05	<0.001	−0.20 ± 0.03	<0.001
Perceived impact of myotonia			−1.64 ± 0.48	0.001	−1.63 ± 0.46	<0.001	−1.43 ± 0.46	<0.01	−1.51 ± 0.42	<0.001
Memory			−0.59 ± 0.20	<0.01	−0.58 ± 0.19	<0.01	−0.59 ± 0.18	0.001	−0.53 ± 0.17	<0.01
Perception of equal opportunities and political orientations as obstacle	0.28 ± 0.09	0.001	0.27 ± 0.09	<0.01	0.28 ± 0.08	<0.01				
Sex × grip strength					0.11 ± 0.05	0.02	0.10 ± 0.03	<0.001		

The potential independent variables tested with stepwise strategy but not retained in the model are not shown for greater clarity.
[a]Three participants indicated all recreation activities as "not applicable" to them.

no other longitudinal study addressed prediction of participation in slowly progressive neuromuscular disorders. Morley's et al. cross-sectional study identified that a superior health status regarding physical and social functioning was associated with routine activities for people with motor neurone disease, in addition to pain for multiple sclerosis or to emotional well-being for Parkinson's disease [48]. Although pain and emotional well-being were not retained as predictors of participation, associations with physical and social functioning were consistent with our findings. Similarly to our results, the cross-sectional analysis of our baseline sample identified among other family income, lower strength, or greater fatigue as associated factors of participation restriction specifically in housing, mobility, and recreation categories. At that time, Gagnon et al., however, identified mainly different environmental factors, such as perceiving government and public services, social support and attitudes of family and friends, and technology as obstacles. Those models explained higher percentages of variance but were built with logistic regression models, a different analysis strategy, and no control for potential confounding variables. Our theoretical models identified apathy as a potential predictor of participation. However, as it was not recorded at baseline in our study, we may have missed a significant predictor

Table 13. Summary of predictors by category of participation.

	Global participation	Daily activities	Nutrition	Fitness	Personal care	Housing	Mobility	Social activities	Community life	Recreation
Personal factors										
CTG repeats expansion size			X							
Family income								X		X
Body mass index	X	X					X			
Walking distance						X			X	
Time to stand and walk	X	X	X		X					
Grip strength	X	X			X		X	X		X
Perceived impact of myotonia	X	X					X			X
Memory										X
Forced vital capacity				X						
Fatigue				X		X				
Functional independence for bowel management			X							
Environmental factors										
Not living at ground level						X				
Use of community services of adapted transportation	X	X					X			
Perception of physical environment and accessibility										
Obstacle	X	X				X		X	X	
Facilitator					X					
Perception of technology as facilitator					X					
Perception of equal opportunities and political orientations as obstacle										X
Sex × grip strength								X		X

Age, sex, phenotype, and education were always controlled for.

of participation restriction considering the work of Van Heugten et al., who found a significant association between apathy and participation restriction [49]. Participation restriction in the current study has been predicted by slightly different variables compared to health-related quality of life, an associated concept. Indeed, lower health-related quality of life was found to be significantly predicted in DM1 by higher age, poorer acceptance of the illness, greater level of depressive symptoms [50], lower education, higher fatigue [51], and severe muscular impairment, no employment, specific personality traits, endocrine and metabolic abnormalities, participation dissatisfaction, and higher daytime sleepiness [17]. Due to a more biomedical approach, environmental factors were also not considered in those studies. The present study supports that environmental factors predict participation 9-year later and should be assessed by rehabilitation professionals.

Implications for clinical practice

Predictors can be used to optimize the annual evaluation of participation recommended by Ashizawa et al. [28] and help to target specific interventions. The usefulness of these predictors can be highlighted in regard to daily and social activities. The impact of CTG repeat expansion size on participation restriction is also important to regard specifically.

Daily activities

Personal predictors of participation in daily activities identified in the current study are mostly aligned with the five known more prevalent and impairing symptoms of DM1 (in order of importance): muscle weakness, fatigue, daytime sleepiness, myotonia, and balance issues [12,52]. As lower grip strength and not perceiving impact of myotonia in daily living were often found at the same time as predictors, it is possible that greater weakness leads to the impossibility for adults with DM1 to perceive myotonia, which could predict higher participation restriction over time. Other predictors were, however, distinctive, such as BMI, functional independence for bowel management, or forced vital capacity. Higher BMI was also found to predict higher daytime sleepiness [53] which has important effects on quality of life [17]. Recently in DM1 population issues with bowel control have been found to touch more than two out of three individuals (68.4%) with many of them who had reported having to make lifestyles change because of fecal incontinence [54]. As respiratory impairment is the leading cause of death in DM1 adults [55,56], forced vital capacity, not surprisingly, predicted participation restriction. Intellectual capabilities were however unexpectedly not retained as predictors. Despite the importance of cognitive functioning on daily activities among the aging population [57], as intellectual capabilities in DM1 adults are often barely normal [58], they might play a minor role in participation restriction. However, since the level of education was used in the modeling, a moderate positive association between the level of education and intellectual capabilities might explain why they were not retained as predictors. In the present study, the chosen variables might also be less representative compare to a measure of concrete performance or a composite score with multiple indicators.

- For rehabilitation professionals, this emphasizes the need to assess exhaustively personal factors of adults with DM1 to better detect potential participation restriction over time and not only the most prevalent symptom.
- The use of indicators could help rehabilitation professionals monitor efficiently and accurately disease progression. For example, specific measurement tools link to some of the predictors identified in the present study had been recommended during international workshops [59–61] (e.g., Timed-up and go, QMT, and dynamometer). Measurement tools in this study may nevertheless differ (e.g., walking capacity with the 10-meter walk recommended test instead of the 2-minute walk test).
- Some predictors linked to the disease could be positively modified by rehabilitation. For example, moderate-intensity strength-training program had been found to had no adverse effects in DM1 [62] and, with modest evidence, to induce

strength and physical capacity improvements [63]. Cognitive behavioral therapy had also been reported as a promising intervention to manage fatigue in this population [64].

Regarding the environmental factors, perception of obstacle in physical environment and accessibility was a recurrent predictor. Another study found that obstacles in physical environment and accessibility limit adults with DM1 to access and navigate in the community, because of narrow aisles and poor condition of sidewalks [65]. Not living at ground level, using community services of adapted transportation, and perceiving physical environment and accessibility, and technology as facilitators were also predictors of participation restriction over time in daily activities. Although, half of the cohort lived at the same floor during study, those who stayed at a higher level were more likely to move to a ground level, suggesting occurrence of architectural barriers over time. When staying at higher level, more participation restrictions over time are expected for housing category. Also, adults with DM1 who use community services and perceive facilitators in their environment (e.g., using technology to facilitate their activities), are more likely to have severe impairments and thus higher participation restriction over time.

- During the evaluation process, rehabilitation professionals could assess specifically those environmental factors to better identify adults at risk of having higher participation restriction in daily activities over time.
- Using a prognostic approach, rehabilitation professional could plan to intervene on environmental factors during the continuum of care. Indeed, it is possible to foresee potential obstacles (e.g., avoiding second-floor apartment when moving) or delay in resources allocation (e.g., discussing home adaptation program several months before needed as part of typical plan to manage DM1) [19].

Social activities

Social activities domain and recreation were predicted by distinctive indicators. They were the only categories where family income predicted participation restriction over time. One qualitative study in DM1 also identified that lower financial resources hinder participation specifically in recreational activities [65]. As social assistance is available in Québec (Canada), financial resources could be enough to provide the needs in essential activities related to daily activities (e.g., nutrition, housing, mobility) but not in social activities (e.g., recreation). However, social activities theoretically provide more opportunities to bind with other people. Social connections allow to receive support from relatives and find a sense of cohesion in the society (i.e., sense of trust and reciprocity with the wider community) which are milestones to achieve successful aging and better health [66]. Recently, a Canadian longitudinal study suggested a formal causal relationship between family income and self-rated health [67], supporting the importance to consider income as a predictive factor of participation. In addition, memory and perception of equal opportunities and political orientations as obstacle were also distinctive predictors of poorer recreation over time. These specific predictors should be addressed by rehabilitation professionals and policy makers.

- For personal factors, family income and memory should be considered by rehabilitation professionals when assessing long-term social activities participation to target more specific interventions. For example, after making sure they received all allowances they are eligible for, individuals could be invited to review the proportion of the budget allocated to social and recreational activities and be directed to free or lower-cost local leisure's services.
- For environmental factors, rehabilitation professionals could specifically assess the perception of opportunities of DM1 people to identify adults further at risk of restrictions. A deeper assessment of the facilitators and obstacles should follow to target the most important perceived obstacles in a long-term intervention plan. As perceived obstacles could be related to community or society environment in addition to personal context, along with the prognostic approach, a Community-based rehabilitation (CBR) approach [68] might be useful for rehabilitation professional and stakeholders. The CBR approach consist of enabling rehabilitation professional to organize the delivery of rehabilitation services in the community and the stakeholders to build a consistent policy and management strategy across health, education, livelihood, and social systems. For examples, to optimize social activities, rehabilitation professionals could give specific coaching on advocating to caregivers of DM1 individual, which may be a role they seek when attending to the clinic [69], and stakeholders could increase community programs offering equal opportunities in a coherent services delivery [29].

For social activities domain and recreation, the contribution of sex to the prediction of participation restriction over time was modified by interaction between sex and grip strength. Such an interaction is complex to explain. To understand the impact of gender on participation restriction over time according to the strength, further study using sex versus gender roles identification and raw and percentage of the predicted value for grip strength would be necessary. For community life, our study might, however, have failed to capture more personal and environmental factors predicting participation restriction as it was the category with the lowest level of explained variance. Further studies could take an interest in better documenting community life restriction considering its importance for social connections.

CTG repeats expansion size and participation

Along with the work of Cumming et al., the current study found that CTG repeats expansion size predicted accomplishment level of participation when used as a single predictor [70]. When considering other personal and environmental factors, CTG repeats expansion size only predicted participation in nutrition. Even if CTG repeats expansion size constitutes a marker of disease severity related to impairments, including muscular weakness [70,71], fatigue [53] or restrictive respiratory syndrome [72], and socially deprived situation [18], it is not completely surprising that it weakly predicts participation. CTG repeats expansion size are not always clinically significant, nor predictive of disease severity from an individual standpoint [73]. In recent years, participation restriction is considered as a social product resulting from a disrupted interaction of the person with his environment [23]. In fact, participation restriction could not solely be attributed to personal factors, such as genetic. Many studies documented the multifactorial nature and importance of environmental factors in the onset of participation restriction for various populations, including DM1 [e.g., 39,74–78], and our study supports this evidence.

- By better informing adults with DM1 and their relatives and reducing the perception of "fatality" associated with having a progressive genetic disease [79], rehabilitation professionals may give people hope of being able to increase their partici-

pation. Such knowledge might also help them to engage more actively in their care and address the modifiable personal and environmental factors influencing their participation restriction.

Study strengths and limits

This study identified predictors of long-term participation restriction with an important cohort of adults with DM1 considering a comprehensive set of variables. The analysis strategy was based on theoretical models from the literature review and an interdisciplinary perspective, and with a three-step process to increase the stability of the identified predictors. The study nevertheless has some limits. First, as the cohort decreased by 43% (of which 69% deceased) between baseline and follow-up, predictors identified might differ for adults more severely affected with DM1. At baseline, there were a substantial number of refusals largely because of lack of interest, a symptom (i.e., apathy) generally related to DM1 [80]. Severely affected adults might thus be underrepresented. Distribution of CTG repeats expansion size and proportion of phenotype were however reported to be similar between participants and non-participants at baseline [40] and at follow-up. Second, although there was only one participant with outliers data excluded prior to the analysis, the heterogeneity occasioned by including persons with different disease duration can be a limitation. It was impossible to control for disease duration in the regression models due to the presence of missing values. We nevertheless systematically controlled for age and phenotype which the latter considers both age at symptom onset and CTG repeat expansion size, leading to an indirect control for disease duration. As different disease duration reflects the reality of this complex and heterogeneous disease, we think it increased the potential of generalization of the predictors regardless of the stage of the illness. Our cohort is however potentially composed of more women with less severely affected disease in the context of universal healthcare system and several social programs. Third, as expected, statistical power did not allow us to identify all possible predictors, but only the stronger ones (i.e., effect size >0.2). Already controlling for four potential confounding variables, the addition of more than seven independent variables to the final regression models was leading to an increase of type II error. Smaller predictors such as intellectual capabilities were thus potentially unidentified. Fourth, as it was a secondary analysis, some variables were used as a proxy (e.g., ankle dorsiflexors as a proxy for lower limb strength) or not available (e.g., apathy) which could have led to sub-optimal predictors or a lower percentage of explained variance for statistical models. For example, as it was recently documented that knee extensors muscle group was more significant for activity and participation in DM1 than ankle dorsiflexors [81], these muscle groups would have been more relevant to identify adults who are at risk of participation restriction. Still, the relative importance of predictors was difficult to estimate due to the multiple imputations which prevent the use of standardized coefficients. In addition, only a few objective environmental factors were documented in the study. For example, our study might have failed to capture the environmental factors predicting participation in nutrition and fitness. Qualitative design studies may help to pinpoint particularities in environmental factors and to provide a more detailed explanation of how participation restriction occurred over time. Inclusion of the people with DM1 and their relatives in such study could also add important information, notably when the theoretical model is revised by a team. Fifth, considering the possible diminished disease awareness in DM1 population [82] and as participation questionnaire and some measurements were self-reported, desirability bias is possible. To limit this bias, participants were encouraged to respond honestly. Finally, predictors for long-term education and employment restriction were not assessed [27] and, acknowledging their importance in adulthood, further studies should consider them.

Conclusions

This study identified predictors of long-term participation restriction. Such predictors could optimize the evaluation and intervention process in order to implement better prognostic approach. It might allow to identify adults at risk of having higher participation restriction over time and offer opportunities to improve the long-term management of the disease by targeting specific interventions (e.g., moderate-intensity exercise, cognitive behavioral therapy, or community-based approach). Family income, BMI, walking distance, time to stand and walk, grip strength, perceived impact of myotonia in daily living, and fatigue were the most found predictors for personal factors. For environmental factors, using community services of adapted transportation and perception of obstacles in physical environment and accessibility were the most found predictors. The majority of those predictors may be assessed in a clinical settings and be positively modified by rehabilitation and promising environmental solutions or policy change, such as targeting universal community accessibility in physical environment. Rehabilitation professionals could improve long-term management for more at risk adults with DM1 by using an interdisciplinary and community-based intervention plan targeting those personal and environmental factors. Further research is, however, still needed to confirm the present results as well as to clarify the associations between personal and environmental factors, on the one hand, and long-term participation restriction for this population, on the other.

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