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

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Purpose: For slow progressive neuromuscular disease, anticipatory guidance and long-term monitoring of participation is a crucial part of rehabilitation services. To improve anticipatory guidance, professionals must identify adults 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 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.

Keywords: health services; longitudinal studies; myotonic dystrophy; patient care management; rehabilitation; social participation.

Implications for rehabilitation:

- To implement better anticipatory guidance for rehabilitation services, predictors could better inform rehabilitation professional to recognize individuals at risk of higher participation restriction over time, to target specific interventions and to bring policy change.
- 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.
- For personal factors, predictors to monitor are in order of importance: grip strength, time to stand and walk, perceived impact of myotonia in daily living, body mass index, fatigue, family income, walking distance, CTG expansion size, memory, forced vital capacity, and bowel management.
- For environmental factors, predictors to monitor are in order of importance: perception of obstacle in physical environment and accessibility, use of community services of adapted transportation, not living at ground level, perception of obstacle in equal opportunities and political orientations, and perception of facilitator in physical environment and accessibility or in technology.

Introduction

Worldwide, myotonic dystrophy type 1 (DM1) is the most prevalent adult-onset form of muscular dystrophies [1]. DM1 touches 5 to 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) lead 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, 10, 11], DM1 implies heterogeneous symptoms and impairments [1]. While one individual may experience progressive distal to proximal weakness, fatigue, excessive daytime sleepiness, myotonia (i.e. delay in relaxing muscle after voluntary contraction), pain [12], apathy, depression, and personality disorder [13], another may have, in addition to weakness, the presence of cataracts, cardiac and respiratory impairments, digestive and endocrine deficits [14] and mild cognitive impairments (e.g. attention, visuospatial and constructional disabilities and dysexecutive syndrome) [15]. Adults with DM1 often live in deprived social environment with lower income, education, and support [16, 17], including limited access to social and medical services [18, 19]. Those personal and environmental factors are likely to lead to disability, which involve participation restriction [20].

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 [21]. To guide policy development and services delivery, this model conceptualised participation restriction as a result of a

disruptive interaction over time of personal and environmental factors [22]. Apart from the consequences in themselves 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 [20]. Associated with quality of life [16, 23], participation restriction was reported as a predictor of reduced cognitive functioning [24] and greater risk of mortality [25] in general aging population. As DM1 is a slow progressive neuromuscular disease, participation restriction gradually increase over time. A clinically significant increase of 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%) [26]. In order to optimize participation of adults with DM1, medical and rehabilitation long-term follow-up is therefore needed.

Long-term monitoring should be a determinant part of rehabilitation services for adults with DM1 [27]. Follow-up and referrals have, however, been often reported as fragmented or happening too late in the process [28, 29, 30, 31]. In addition, unmet needs for social care and rehabilitation services are reported in a large proportion of adults with DM1 (68-82%) [32] and their relatives (19-31%) [31]. Since 2010, anticipatory guidance and health supervision have been advocated to help adults with DM1 in the disease management and optimisation of their participation [18]. Annual evaluation of participation in daily living and social activities is also recommended [27]. To implement better anticipatory guidance 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. Due to the dominant inheritance, social environment of adults with DM1 often implies low social support from family members [16] and a high caregiver burden as he/she often has to look after more than one adult with DM1 [31]. Yet, even if environmental factors played a role in the presence of participation restriction, they are sparsely studied in neuromuscular research, and knowledge gap is still present. This study thus aimed to identify personal and environmental predictors of participation restriction over nine years in adults with DM1. This paper extend our previous study describing changes in participation over 9-year [26]. Based on the same participant's sample, predictors were specifically identified for the categories of participation that restriction have been found to clinically significantly increase in our previous study [26].

Method

Design

A longitudinal study comparing baseline (2002-04) and follow-up data (2011-13) 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. Further details on the procedures for patients' selection can be found elsewhere [26, 33, 34, 35]. The study was approved by the Ethics Review Board of the Centre intégré universitaire de santé et de services sociaux du Saguenay–Lac-St-Jean (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 from 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.

[Please insert figure 1 about here]

Variables

To identify the independent variables that could potentially predict participation restriction at the follow-up (dependent variables), theoretical models were built. To do so, an overview of the scientific literature was carried out by the first author and revised

independently by two co-authors. Medline, CINHAL, EMB Reviews, Scopus, and Ageline were searched from 1991 (first publication of the HDM-DCP framework) to 2016. Eligible studies were identified if they satisfied the following criteria: 1- DM1 or normal aging populations; 2-identified predictors of participation or activities of daily living or instrumental activities of daily living or community integration or loneliness; 3-longitudinal studies (cross-sectional and qualitative studies were accepted for studies with DM1 patients only) and 4-English or French articles. Studies were excluded if based on simple correlational analysis. In this secondary analysis, theoretical models for factors predicting participation for each category were built based on data extraction. Then, an interdisciplinary team (psychologist, physiotherapist, biologist, social worker, and research professional specialised in muscular disease) reviewed the theoretical models to add or remove potential predictors in order to better represent DM1 symptomatology. Theoretical models are presented for the seven categories of participation (supplementary table S1). The theoretical models led to the identification of 56 potential independent variables among the personal and environmental factors (figure 1).

Personal and environmental factors (independent variables)

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). Definition of variables and psychometric proprieties of measurement tools are presented in supplemental appendix (supplementary file).

Participation restriction (dependent variables)

Participation restriction was assessed with the short 3.1 version of the Assessment of Life Habits Questionnaire (LIFE-H) [36]. This questionnaire includes 77 activities covering 12 categories of participation (number of activities) divided in two domains: 1- daily activities: nutrition (4), fitness (4), personal care (8), communication (8), housing (8), and mobility (5); 2- social activities: responsibilities (8), interpersonal relationships (7), community life (8), education (2), employment (8), and recreation (7). Based on self-reported accomplishment level in 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 good 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) [37]. Minimal clinically important difference (MCID) was considered with a change of 0.5 (/9) [38].

Data analysis

Participant characteristics were shown with mean (SD) for continuous variables and frequency (%) for categorical variables. Among the 56, eight independent variables presented missing data of less than 15% that were accommodated at the composite score [39] with five multiple imputations as recommended by Graham (2007) [40]. Multiple

imputation is a reliable method which simulates multiple possible scenarios approximating the missing data [41]. Disease duration, BBS, and NEO-FFI were excluded due to presence of more than 15% missing data. Nominal variables were coded as dummy; and ordinal variables of six categories and more were considered as continuous variables. Multivariate linear regression was used to predict participation restriction after a 9-year period with independent variables at baseline. As 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 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 composing category of participation (data not shown). All variables with alpha $\geq$ 0.10 were excluded from the subsequent analysis regarding the exploratory process [42]. 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 S2-11). Hierarchical method consisted of selecting first the potential personal and environmental factors with alpha < 0.10 and then ordering variables that correlated more strongly with participation restriction (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 [26, 33, 43], 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 graphical review of standardized predicted values on studentized residuals as well as with Kolmogorov-Smirnov analysis. Multicollinearity were also tested with variance inflation factor. The statistical analyses were performed using SPSS software (version 25.0 for Windows) and an alpha of 0.01 was used for significance of the final multivariate regression models as several were performed.

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 likely caused by uncontrolled diabetes. Compared to the 114 who participated in follow-up, the 85 adults who did not participate to follow-up did not differ for sex, CTG repeats and phenotypic distribution. Yet, they were older ($p<0.01$) and had less participation restriction ($p<0.01$). Aged between 20 and 77 years at baseline, participants were mostly women and with adult phenotype (table 1). 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, only four were using technical aids (can or roller walker, data not shown). 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. 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.

[Please insert figure 2 and table 1 about here]

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 2). 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. Little meaningfully changes in coefficient estimates were observed for a few variables (e.g. perceived impact of myotonia: -0.47 to -0.51) when removing the confounding variables.

[Please insert table 2 about here]

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 3). 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 3).

[Please insert table 3 about here]

For all daily activities' category, each model explained between 30% and 49% of the variance of participation restriction at follow-up (tables 4-8).

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 4). However, a lower forced vital capacity and a higher fatigue predicted greater participation restriction in fitness (table 5). 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.

[Please insert tables 4 and 5 about here]

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 6).

[Please insert table 6 about here]

Housing. After controlling for confounding variables, a smaller walking distance and a higher fatigue, not living at ground level, and perception of physical environment as obstacle predicted greater participation restriction in housing (table 7).

[Please insert table 7 about here]

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 8).

[Please insert table 8 about here]

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 8). 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 9). 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.

[Please insert table 9 about here]

For community life and recreation category, model explained respectively 20% and 52% of the variance of participation restriction at follow-up (tables 10 and 11).

Community life. A smaller walking distance and perception of physical environment as obstacle predicted greater participation restriction in community life (table 10). However,

community life was the category with the lowest level of explained variance.

[Please insert table 10 about here]

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 11). In addition, sex interacted with grip strength by modifying the effect of sex the same way did the social activities domain.

[Please insert table 11 about here]

Discussion

This study identified personal and environmental predictors of participation restriction over a 9-year period. Predictors slightly differ between global participation, daily activities domain and social activities domains. For personal factors, predictors of participation restriction were in order of importance (# presence in all final models): lower grip strength (6), longer time to stand and walk (4), perceiving impact of myotonia in daily living (4), higher BMI (3), greater fatigue (2), lower family income (2), smaller walking distance (2), higher CTG expansion size (1), lower memory (1), lower forced vital capacity (1), and lower functional independence for bowel management (1). For environmental factors, predictors were in order of importance: perception of obstacle in physical environment and accessibility (5), use of community services of adapted transportation (3), not living at ground level (1), perception of obstacle in equal opportunities and political orientations (1), and perception of facilitator in physical environment and accessibility (1) or in technology (1). Similarly to our results, the cross-

sectional analysis of our baseline sample identified among other family income, lower strength or greater fatigue as predictors of participation restriction specifically in housing, mobility, and recreation categories [44]. At that time, Gagnon et al. (2008), however, identified mainly different environmental predictors, 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 of participation restriction considering the work of Van Heugten et al. (2018), who found significant association between apathy and participation restriction [45]. To our knowledge, no other study identified predictors of participation restriction in DM1 with cross-sectional nor longitudinal studies. Nevertheless, participation restriction in the current study has been predicted by slightly different variables compare 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 [46], lower education, higher fatigue [47], and severe muscular impairment, no employment, specific personality traits, endocrine and metabolic abnormalities, participation dissatisfaction, and higher daytime sleepiness [16].

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 [31, 48]. 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. Recently in DM1 population issues with bowel control has 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 faecal incontinence [49]. Higher BMI was also found to predict higher daytime sleepiness which has important effects on quality of life [16]. For rehabilitation professionals, this emphasizes the need to assess exhaustively personal factors of adults with DM1 to better detect potential participation restriction over time. Regarding the environmental factors, 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 [50]. 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. Adults with DM1 who use community services and perceive facilitators in their environment (i.e. using for example technology to facilitate their activities), are more likely to have severe impairments and thus higher participation restriction over time. Rehabilitation professionals could assess those

environmental factors to better identify adults at risk of having higher participation restriction in daily activities over time.

Social activities

Social activities domain and recreation 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 in recreational activities [50]. As social assistance is available in Québec, 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 [51]. Considering that perception of equal opportunities and political orientations as obstacle was also a predictor of participation restriction over time, poorer social activities and recreation should be addressed by rehabilitation professionals and policy makers (e.g. by the reappraisal of the financial assistance policies). 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 being a woman or a man on participation restriction over time according to the strength, further study using a gender roles identification and raw and percentage of 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 study could take an interest in better documenting community life restriction considering its importance for social connections.

Implications for clinical practice

Better detection of adults with increased risk of participation restriction

In view of the present results, rehabilitation professionals could better detect current and future needs of adults with DM1 with the use of objective indicators, such as time to stand and walk, grip strength, or BMI. Predictors could also be used to optimize the annual evaluation of participation in daily living and social activities recommended by Ashizawa et al. (2018) [27] and to target more specific interventions to optimize participation in the future, such as bowel control or BMI. Attention should be given to reduce obstacles in physical environment and accessibility as well as to increase community services offer. Even if significant improvements over the last decades had been made, better community access, coherent service delivery, technology development, and disability-related socioeconomic policies are promising environmental solutions to promote optimal participation [52]. Further attention is needed to fully understand the impact of environmental factors on participation, such as with qualitative design study.

CTG repeats expansion size and participation

Along with the work of Cumming (2019), the current study found that CTG repeats expansion size predicted accomplishment level of participation when use as a single

predictor [53]. 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 [53, 54], fatigue [35] or restrictive respiratory syndrome [55], and social deprived situation [17], it is not completely surprising that it weakly predicts participation. In recent years, participation restriction is considered as a social product resulting from a disrupted interaction of the person with his environment [22]. 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 onset of participation restriction for various populations, including DM1 [44, 56, 57, 58, 59, 60], 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 [61], rehabilitation professionals may give people hope of being able to increase their participation. 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 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 decrease by 43%

(of which 69% deceased) between baseline and follow-up, predictors identified might differed for adults more severely affected with DM1. Second, statistical power did not allow us to identify all possible predictors, but only the stronger ones. Already controlling for four potential confounding variables, the addition of more than six independent variables to the regression models was leading to an increase of type II error. Third, as it was a secondary analysis, some variables were used as 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 lower percentage of explained variance for statistical models. For example, as it was recently found that knee extensors muscle group was more significant for activity and participation in DM1 than ankle dorsiflexors [62], these muscle group would have been more relevant to identify adults who are at risk of participation restriction. In addition, only 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. Finally, predictors for long-term education and employment restriction were not assessed [26] and, acknowledging their importance in adulthood, further study should consider them.

Conclusion

This study identified predictors of long-term participation restriction. Such predictors could optimize the evaluation and intervention process in order to implement better anticipatory guidance. 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. Ultimately, a better long-term management could be an effective way to diminish disability situation of adults with DM1. Family income, BMI, walking distance, time to stand and walk, grip strength, perceive 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 or facilitator in physical environment and accessibility were the most found predictors. The majority of those predictors may be positively modified by rehabilitation and promising environmental solutions or policy change, such as targeting universal community accessibility in physical environment. 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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Disclosures

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List of abbreviations

2MWT: 2-Minute Walk Test. BBS: Berg Balance Scale. BMI: body-mass index. CTG: cytosine-thymine-guanine. CVLT: California Verbal Learning Test. DM1: myotonic dystrophy type 1. DSS: daytime sleepiness scale. FIM: Functional Independence Measure. HDM-DCP: Human Development Model - Disability Creation Process. ICC: intra-class correlation coefficient. IT: interdisciplinary team suggestions to complete theoretical models. JAMAR: Jamar Dynamometer. KFSS: Krupp Fatigue Severity Scale. LIFE-H: Assessment of Life Habits Questionnaire. MCID: Minimal clinically important difference. MQE: Measure of the Quality of the Environment. NEO-FFI: NEO Five-Factor Inventory. QMT: quantitative muscular testing. RSES: Rosenberg Self-Esteem Scale. SCL-90: Symptom Checklist-90 Revised. SCWT: Stroop Color and Word Test. TUG: Timed-up and Go. WAIS-R: Wechsler Adult Intelligence Scale-Revised.

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Table 1. Characteristics of DM1 participants (n=114)

Personal factors	Baseline	Follow-up
Identity factors		
Age (y)	43.5 (10.4)	52.2 (10.3)
Sex (women = 0)	72 (63.2)	
Education (y)		
≤ 11	50 (43.9)	50 (43.9)
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)
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)
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	19.9 (8.1) [3-38]	
Missing data, n (%)	*33 (28.9)*	
CTG repeats expansion size	777 (516)	923 (505)
Missing data, n (%)		*2 (1.8)*
Number of comorbidity	1.8 (1.6)	-
Body mass index	25.1 (5.4)	25.7 (5.9)
Literacy for filing form		
No help needed	93 (81.6)	87 (76.3)
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)
Recent life stressors (no = 0)	101 (88.6)	-
Active smoking (yes = 1)	37 (32.4)	28 (24.6)
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)
Missing data, n (%)		16 (14.0)
Ankle dorsiflexors strength (QMT, *Nm*)	16.4 (7.8)	7.4 (4.8)
Missing data, n (%)	*8 (7.0)*	*7 (6.1)*
Dominant grip strength (JAMAR, *Kg*)	12.2 (10.1)	10.9 (9.2)
Missing data, n (%)	1 (0.9)	4 (3.5)
Perceived impact of myotonia (yes = 1)	46 (40.4)	-

Balance (BBS, /56)	52.9 (6.8)	47.1 (13.6)
Missing data, n (%)	*30 (26.3)*	*7 (6.1)*
Intellectual capabilities		
Intellectual quotient (WAIS-R)	83.1 (8.4)	84.9 (9.0)
Missing data, n (%)	*3 (2.6)*	*2 (1.8)*
Executive function (SCWT, T-score)	46.9 (6.9)	49.9 (7.5)
Missing data, n (%)	*5 (4.4)*	*4 (3.5)*
Memory (CVLT, Z-score)	0.19 (1.1)	-0.5 (1.2)
Missing data, n (%)		*1 (0.9)*
Breathing capabilities		
Forced vital capacity (Spirometer, *in Liter*)	3.1 (0.9)	2.7 (0.9)
Missing data, n (%)	*15 (13.2)*	*15 (13.2)*
Protection and resistance capabilities		
Frequency of physical activity		
Never	39 (34.2)	62 (54.4)
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)
Missing data, n (%)		*2 (1.8)*
Fatigue (KFSS, /63)	39.1 (15.3)	43.4 (15.4)
Missing data, n (%)		*1 (0.9)*
Actual health self-assessment		
Bad to passable	30 (26.3)	44 (38.6)
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)
Missing data, n (%)	*11 (9.6)*	*1 (0.9)*
Anxiety symptoms (SCL-90, T-score)	46.6 (9.3)	46.2 (8.8)
Missing data, n (%)	*10 (8.8)*	*1 (0.9)*
Self-esteem (Rosenberg, /40)	31.0 (4.5)	30.2 (5.2)
Missing data, n (%)	*1 (0.9)*	
Personality traits (NEO-FFI, T-score)		
Neuroticism	48.2 (9.3)	50.6 (10.2)
Extraversion	50.0 (8.6)	47.8 (9.8)
Openness	41.3 (7.5)	41.3 (8.2)
Agreeableness	49.2 (9.4)	48.0 (9.0)
Concientiousness	50.3 (8.7)	46.5 (8.1)
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)
Environmental factors	Baseline	Follow-up
Personal context		
Living arrangement (home alone = 0)	18 (15.8)	33 (28.9)
Food insecurity (no)	96 (84.2)	99 (86.8)
Floor of living area (ground level = 0)	37 (32.5)	71 (62.3)
Use of community services (yes = 1)		
Meals delivery	15 (13.2)	8 (7.0)
Household assistance	50 (43.9)	56 (49.1)
Adapted transportation	21 (18.4)	26 (22.8)
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)	Baseline	Follow-up
Global participation	-8.2 (0.8)	-7.7 (1.2)*
Daily activities	-8.2 (0.8)	-7.8 (1.2)
Nutrition (n=112) [1]	-8.6 (0.8)	-7.8 (1.8)*
Fitness	-8.1 (1.2)	-7.1 (1.7)*
Personal care	-8.7 (0.5)	-8.0 (1.4)*
Housing	-7.5 (1.5)	-7.4 (1.5)
Mobility	-7.5 (1.7)	-7.0 (2.1)*
Social activities	-8.2 (0.9)	-7.7 (1.4)*

Community life	-8.5 (1.0)	-7.7 (2.1)*
Recreation (n=111) [1]	-7.3 (2.5)	-5.8 (3.0)*

Abbreviation (precision on scoring interpretation): 2MWT: 2-Minute Walk Test (a higher score indicated a higher 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 (a higher Z-score indicated a higher 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 (a higher T-score indicated a higher 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 (a higher score indicated a higher symptomatology). SCWT: Stroop Color and Word Test (a higher T-score indicated a higher 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).

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

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

* Change of 0.5 points is clinically significant.

Table 2. Unstandardized Regression Coefficients in the Final Multivariate Model Identifying with Chunkwise Strategy the Best Predictors of Participation Restriction **Globally** (N=114)

	Model 1 (ajusted R^2 = 0.19)		Model 2 (ajusted R^2 = 0.43)		Model 3 (ajusted R^2 = 0.50)		Model without confounding (ajusted R^2 = 0.47)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

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

Table 3. 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 (ajusted R^2 = 0.22)		Model 2 (ajusted R^2 = 0.48)		Model 3 (ajusted R^2 = 0.54)		Model without confounding (ajusted R^2 = 0.50)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

Note. 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 **Nutrition** (N=112)[1]

	Model 1 (ajusted R^2 = 0.14)		Model 2 (ajusted R^2 = 0.42)		Model without confounding (ajusted R^2 = 0.35)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

[1] Two participants indicated all nutrition activities as 'not applicable' to them.

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

Table 5. Unstandardized Regression Coefficients in the Final Multivariate Model Identifying with Chunkwise Strategy the Best Predictors of Participation Restriction in **Fitness** (N=114)

	Model 1 (ajusted R^2 = 0.13)		Model 2 (ajusted R^2 = 0.30)		Model without confounding (ajusted R^2 = 0.14)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

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

Table 6. 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 (ajusted R^2 = 0.14)		Model 2 (ajusted R^2 = 0.41)		Model 3 (ajusted R^2 = 0.49)		Model without confounding (ajusted R^2 = 0.45)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

Note. 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 **Housing** (N=114)

	Model 1 (ajusted R^2 = 0.19)		Model 2 (ajusted R^2 = 0.38)		Model 3 (ajusted R^2 = 0.43)		Model without confounding (ajusted R^2 = 0.37)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

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

Table 8. Unstandardized Regression Coefficients in the Final Multivariate Model Identifying with Chunkwise Strategy the Best Predictors of Participation Restriction in **Mobility** (N=114)

	Model 1 (ajusted R^2 = 0.17)		Model 2 (ajusted R^2 = 0.31)		Model 3 (ajusted R^2 = 0.34)		Model without confounding (ajusted R^2 = 0.34)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

Note. 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 **Social activities** (N=114)

	Model 1 (ajusted R^2 = 0.15)		Model 2 (ajusted R^2 = 0.31)		Model 3 (ajusted R^2 = 0.36)		Model 4 (ajusted R^2 = 0.39)		Model without confounding (ajusted R^2 = 0.37)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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 X grip strength							0.06 ± 0.02	0.02	0.05 ± 0.01	0.001

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

Table 10. 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 (ajusted R^2 = 0.09)		Model 2 (ajusted R^2 = 0.14)		Model 3 (ajusted R^2 = 0.20)		Model without confounding (ajusted R^2 = 0.18)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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

Note. 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 **Recreation** (N=111)[1]

	Model 1 (ajusted R^2 = 0.18)		Model 2 (ajusted R^2 = 0.44)		Model 3 (ajusted R^2 = 0.49)		Model 4 (ajusted R^2 = 0.52)		Model without confounding (ajusted R^2 = 0.52)	
	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	P value	$\beta \pm SE$	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 X grip strength							0.11 ± 0.05	0.02	0.10 ± 0.03	< 0.001

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

[1] Three participants indicated all recreation activities as 'not applicable' to them.

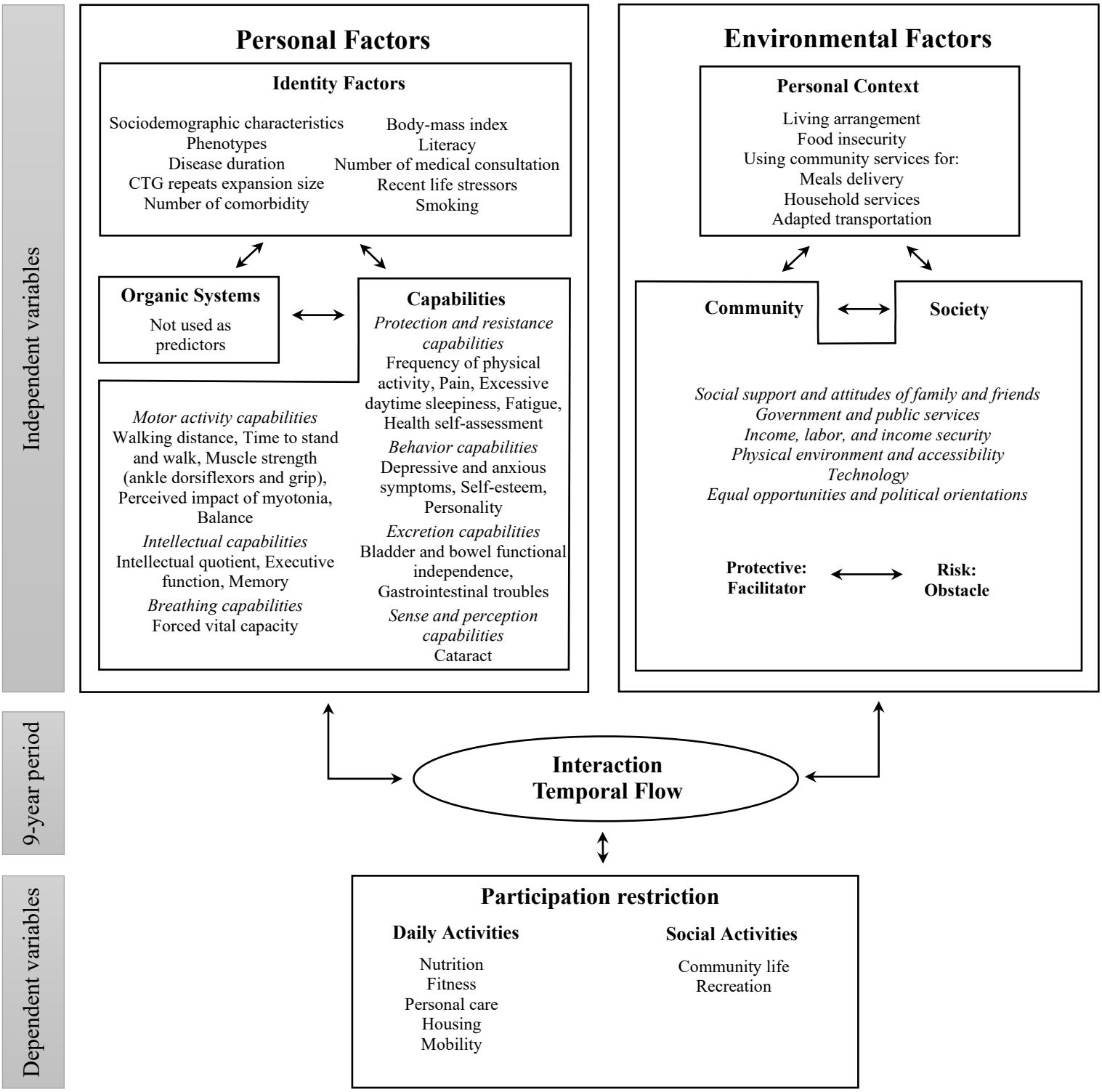
Independent variables
9-year period
Dependent variables
Personal Factors
Identity Factors
Sociodemographic characteristics
Phenotypes
Disease duration
CTG repeats expansion size
Number of comorbidity
Body-mass index
Literacy
Number of medical consultation
Recent life stressors
Smoking
Organic Systems
Not used as predictors
Capabilities
Protection and resistance capabilities
Frequency of physical activity, Pain, Excessive daytime sleepiness, Fatigue, Health self-assessment
Behavior capabilities
Depressive and anxious symptoms, Self-esteem, Personality
Excretion capabilities
Bladder and bowel functional independence, Gastrointestinal troubles
Sense and perception capabilities
Cataract
Motor activity capabilities
Walking distance, Time to stand and walk, Muscle strength (ankle dorsiflexors and grip), Perceived impact of myotonia, Balance
Intellectual capabilities
Intellectual quotient, Executive function, Memory
Breathing capabilities
Forced vital capacity
Environmental Factors
Personal Context
Living arrangement
Food insecurity
Using community services for:
Meals delivery
Household services
Adapted transportation
Community
Society
Social support and attitudes of family and friends
Government and public services
Income, labor, and income security
Physical environment and accessibility
Technology
Equal opportunities and political orientations
Protective: Facilitator
Risk: Obstacle
Interaction
Temporal Flow
Participation restriction
Daily Activities
Nutrition
Fitness
Personal care
Housing
Mobility
Social Activities
Community life
Recreation

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

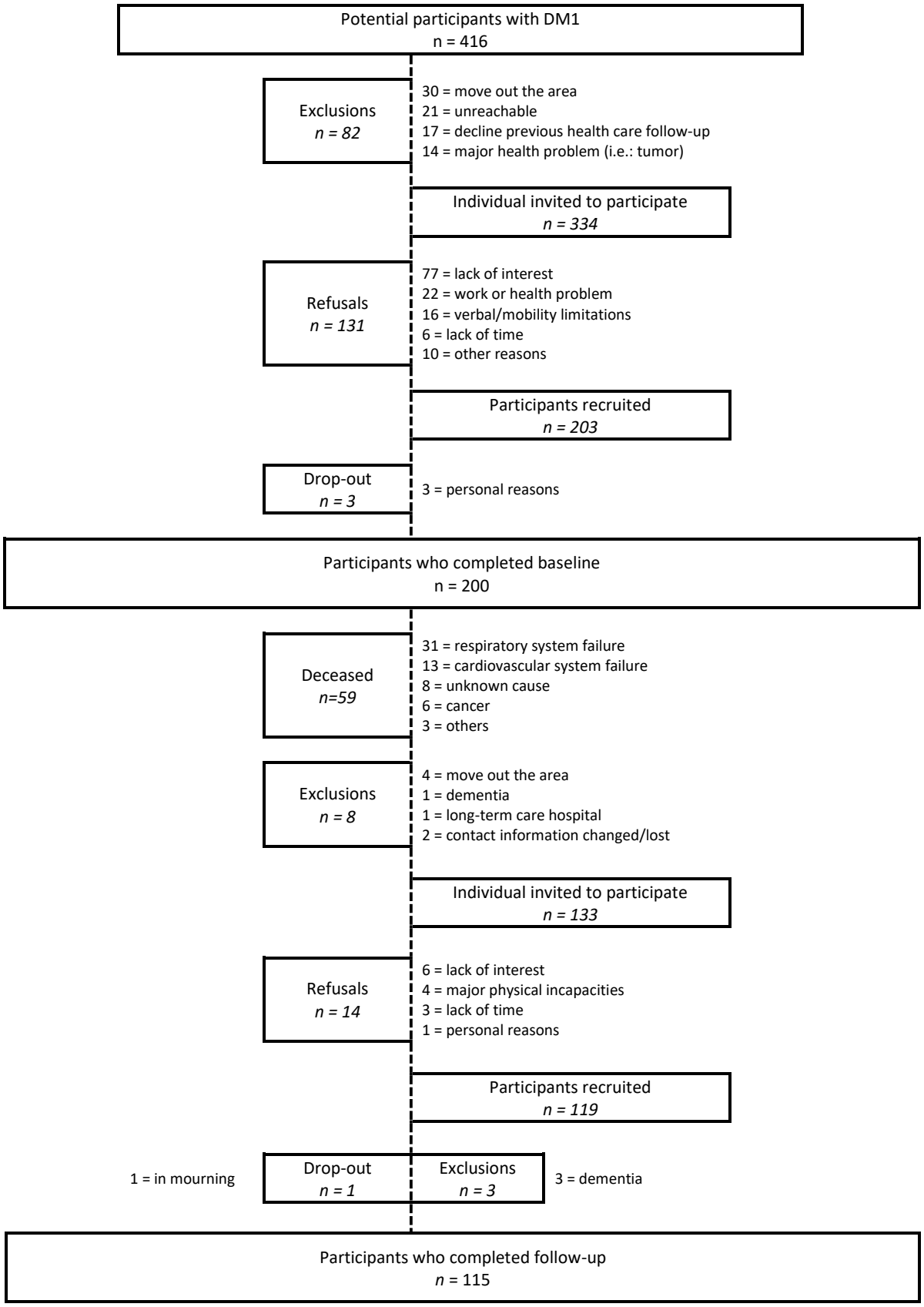
Potential participants with DM1
n = 416
Exclusions
n = 82
30 = move out the area
21 = unreachable
17 = decline previous health care follow-up
14 = major health problem (i.e.: tumor)
Individual invited to participate
n = 334
Refusals
n = 131
77 = lack of interest
22 = work or health problem
16 = verbal/mobility limitations
6 = lack of time
10 = other reasons
Participants recruited
n = 203
Drop-out
n = 3
3 = personal reasons
Participants who completed baseline
n = 200
Deceased
n=59
31 = respiratory system failure
13 = cardiovascular system failure
8 = unknown cause
6 = cancer
3 = others
Exclusions
n = 8
4 = move out the area
1 = dementia
1 = long-term care hospital
2 = contact information changed/lost
Individual invited to participate
n = 133
Refusals
n = 14
6 = lack of interest
4 = major physical incapacities
3 = lack of time
1 = personal reasons
Participants recruited
n = 119
1 = in mourning
Drop-out
n = 1
Exclusions
n = 3
3 = dementia
Participants who completed follow-up
n = 115

Figure 2. Participants' flow chart
Note. Already published in Raymond K, et al. (2019) [26] (reuse permission granted).

Personal factors (independent variables)

Identity factors. Sociodemographic characteristics, including age, sex, education, income, and marital status were obtained from participants using a questionnaire. Phenotypes were used to discriminate between adult (onset < 40 y.o.) and late-onset forms (onset > 40 y.o., and/or < 200 CTG repeats, and/or no muscular impairment to minimal signs). When available, disease duration was documented using medical files. CTG repeats expansion size was documented with peripheral blood samples using a standard procedure [1] at the time of the study. Number of comorbidities was assessed using a questionnaire recording common chronic diseases and often associated with DM1 disease. Body-mass index (BMI) was assessed using a bioimpedance balance in kg/m^2. Defined as the ability to understand, evaluate, use, and engage with written texts to participate in society [2], literacy was assessed using a questionnaire asking about the frequency of help needed to read, understand, and filling out forms with a scale from 0 (never) to 2 (often). The number of medical consultations during the last year was recorded based on a self-reported questionnaire. Recent life stressors (e.g., separation, relocation) during the last year were assessed using a questionnaire and then recoded as 0 (lived no life stressor in the last year) or 1 (lived at least one stressor in last year). Smoking was assessed with a yes/no self-reported question.

Motor activity capabilities. Walking distance was assessed in meter with a 2-Minute Walk Test (2MWT). The 2MWT presents excellent intra-rater reliability (intra-class correlation coefficients (ICC): 0.97) for neuromuscular conditions [3]. Indicator of functional mobility, time to stand and walk was assessed with Timed up and Go (TUG)

[4]. The TUG presents good intra-rater reliability (ICC: 0.83) [5] and responsiveness (area under the curve: 0.8) in DM1 [6]. Quantitative muscle testing (QMT) with a handheld dynamometer (Microfet – 2, Hoggan Health Industries, Salt Lake City, UT) was used to document maximal isometric muscle strength of the ankle dorsiflexors with the mean of two trials in newton-meters using a standardized procedure [7]. The QMT presents excellent intra-rater reliability (R^2: 0.96) and good responsiveness (area under the curve: 0.6) in DM1 [8]. Jamar dynamometer (Asimow Engineering Co., Los Angeles, CA) was used to document grip strength in kilograms with standardized procedure [9]. Jamar presents excellent intra-rater reliability (ICC: 0.98) in DM1 [10]. Perceived impact of myotonia in daily living was assessed with the yes/no self-reported question "do difficulty to relax muscle to release object interfere in your daily living?". Balance was assessed with Berg Balance Scale (BBS) [11]. There are no published reliability studies in DM1 for BBS, but a panel of experts reported a high intra-rater reliability (no data shown) [12] which was also found in various population (ICC: 0.98) [13].

Intellectual capabilities. Intellectual functioning was assessed using the full scale intellectual quotient of the Wechsler Adult Intelligence Scale-Revised (WAIS-R)[14]. Although, there are no published reliability studies in DM1, the WAIS-R was found to have excellent intra-rater reliability (ICC: 0.94) in the general population [14]. Executive function was estimated with the Stroop Color and Word Test (SCWT) which presents acceptable reliability in DM1 [15, 16]; the T-score of inhibition capacity task was specifically chosen, as it requires higher executive functioning to execute the task and is a better indicator of executive functioning. The SCWT presents good intra-rater reliability (ICC: 0.80) in aging population [17]. Memory functioning was estimated with the

California Verbal Learning Test (CVLT)[18], with Z-score standardized for age and sex. Although, there are no published reliability studies in DM1, the CVLT has a good to excellent intra-rater reliability (ICC: 0.82 to 0.90) in the general population [19].

Breathing capabilities. Defined as the maximum amount of air that can be exhaled when blowing out as fast as possible and related to the restrictive respiratory syndrome in DM1 [20], forced vital capacity was assessed in liter with a spirometer using a standardized position [21].

Protection and resistance capabilities. Frequency of physical activity was documented with the self-reported question "how many times have you been physically active for 20 to 30 minutes per session, in free time, in the last 3 months?" with seven possible answers from 1 (never) to 7 (4 times or more a week). Pain was assessed with the yes/no self-reported question "does pain interfere in your daily living?". Excessive daytime sleepiness was assessed with the 5-item Daytime Sleepiness Scale (DSS)[22], DSS was specifically devised for DM1 and presents good reliability (ICC: 0.82) in DM1 [23]. Fatigue was assessed with the Krupp's Fatigue Severity Scale (KFSS) with nine items on a 7-point Likert scale [24]. The KFSS presents good to excellent intra-rater reliability (ICC: 0.88) in DM1[23]. Self-assessed health was evaluated using a question with a 5-point Likert scale from 0 (bad) to 4 (excellent).

Behavior capabilities. Depressive and anxious symptoms were recorded in T-scores with respective subscale of the Symptom Checklist-90-Revised (SCL-90) [25]. Although, there are no published reliability studies in DM1, SCL-90 presented a good to excellent intra-rater reliability (Cronbach alpha: 0.62 to 0.96) in the general population [26]. Self-

esteem was assessed using the Rosenberg Self-esteem Scale (RSES) [27]. Although, there are no published reliability studies in DM1, RSES presents a good intra-rater reliability (ICC: 0.84) in the general population [28]. The NEO Five-Factor Inventory (NEO-FFI) was used to provide a general description of personality traits with five major domains (neuroticism, extraversion, openness to experience, agreeableness, and conscientiousness) [29]. Although, there are no published reliability studies in DM1, the NEO-FFI presents a good to excellent intra-rater reliability (ICC: 0.79 to 0.97) in the general population [30].

Excretion capabilities. Specific bladder and bowel functional independence were assessed using the Functional Independence Measure (FIM) [31]. Although, there are no published reliability studies in DM1, the FIM presents an excellent intra-rater reliability for sphincter control (ICC: 0.98) in neuromuscular population [32]. Presence of gastrointestinal troubles was assessed with a yes/no self-reported question "do gastrointestinal troubles interfere in your daily living?".

Sense and perception capabilities. Having cataracts was assessed with a yes/no self-reported question.

Environmental factors (independent variables)

Personal context. Living arrangement (home alone or living with someone; living at ground level or else) was assessed using a questionnaire. Defined as an inadequate or uncertain access to food [33], food insecurity was documented by a yes/no indicator consisting of three questions on food insecurity (diet's monotony, limited accessibility to

food, and difficulty to give balanced meals to children, when applicable) [34]. Using community services for meal delivery, household services, and adapted transportation during the last 12 months were assessed with yes/no self-reported questions.

Community and society. Self-perceived physical and social environment was documented with the Measure of the Quality of the Environment (MQE) version 2.0 [35], using 109 items divided into six domains (number of items): social support and attitudes of family and friends (14), government and public services (27), income, labor, and income security (15), physical environment and accessibility (38), technology (5), and equal opportunities and political orientations (10). The self-perceived environment is scored on a 7-point Likert scale ranging from -3 (major obstacle) to 3 (major facilitator). To facilitate the interpretation, obstacle scoring was positively reported by inverting the score. Although, there are no published reliability studies in DM1, the MQE presents a moderate to high intra-rater reliability (moderate to high kappas for 57% of the items) in cerebral palsy population [36].

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