

# Multiple case study of changes in participation of adults with myotonic dystrophy type 1 : Importance of redesigning accomplishment and resilience

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**Multiple case study of changes in participation of adults with myotonic dystrophy type 1:  
Importance of redesigning accomplishment and resilience**

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**Highlights**

- Since diagnosis, participation decline is slow, heterogeneous, and insidious.
- Social isolation and restriction in life-space mobility progressively appear.
- Redesigning accomplishment with resilience are key facilitators to participation.
- Misfit between personal and environmental factors restricts participation.
- A syndemic interaction may contribute to participation restrictions.

**ABSTRACT**

This study explored and explained changes in participation post-diagnosis with myotonic dystrophy type 1 from the perspective of six adults, their relatives and nurse case managers. A multiple case study was carried out with these triads (n=18) using semi-structured individual interviews, medical charts, and a participation patient-reported outcome measure. The six cases were built around three women and three men (age: 40-56 years; disease duration: 19-39 years). Their “relatives” were mainly family members. Nurse case managers had done annual follow-ups with all the adults for approximately ten years. Changes in participation were characterized generally by: 1) heterogeneity, 2) insidious increase in restrictions, and more specifically by: 3) redesigning accomplishment, 4) progressive social isolation, 5) restrictions in life-space mobility, and 6) increasingly sedentary activities. Important facilitators of participation were the adult’s resilience, highly meaningful activities, social support, living arrangement, and willingness to use technical aids. Barriers were mostly related to symptoms and a precarious

social network, and were affected by misfit and potential syndemic interactions between personal (e.g., comorbidities) and environmental (e.g., stigma) factors. This study identified key facilitators and barriers and their underlying processes, which should be integrated in the evaluation and intervention framework to optimize participation over time.

### **Keywords**

Activities of daily living, environment, neuromuscular diseases, qualitative research, rehabilitation, social participation, myotonic dystrophy, social isolation

### **Abbreviations**

DM1, myotonic dystrophy type 1. HDM-DCP, Human Development Model – Disability Creation Process. ICC, intraclass correlation coefficients. LIFE-H, Assessment of Life Habits. MCID, minimal clinically important difference.

## **1. INTRODUCTION**

Myotonic dystrophy type 1 (DM1), the most prevalent adult form of neuromuscular disorders, is a dominant autosomal hereditary disease often compared to premature aging (1-4). It is a progressive multisystemic condition (5) that frequently leads to multiple impairments and increased restrictions in participation in daily and social activities (6, 7). Participation could be defined by the level of accomplishment and need for assistance in daily and social activities (8, 9). Although important steps have been made in understanding the pathophysiology of DM1, as yet there is no cure (10, 11).

Managing the symptoms and optimizing participation and quality of life, therefore, are often the main goals of the people directly involved, i.e., adults with DM1, their families, and health professionals. To achieve these goals, experts recommend using a prognostic approach, i.e., health supervision with anticipatory guidance (12), and annual follow-ups for preventive and management interventions (13). Implementing such an approach requires a better understanding of short- and long-term changes in participation as well as potential facilitators and barriers and their underlying processes. Efforts to find a cure with well-designed therapeutic trials or longitudinal study also need a deeper understanding of potential factors affecting the outcomes being studied, such as participation, and control of potential confounding factors (adjust for specific personal or environmental factors in analyses) (14).

For adults with DM1, it is known that participation decreases slowly over the long term and more and more activities are impeded (6, 7, 15, 16). However, little is known about perceived changes in participation over time, i.e., changes in daily and social activities and the sequence in which restrictions occur. In the aging population, participation restrictions usually start with instrumental activities of daily living (e.g., household chores) followed by activities of daily

living (e.g., self-care) and mobility (17). In a literature overview of the population with neuromuscular disorders, only one long-term longitudinal study (18) including among others participants with DM1 aimed to elucidate experiences of living with the disease and its impact on activities over ten years. It was found that more time was needed and new strategies were used to continue doing daily and social activities. Housing, mobility, interpersonal relationships, work, and recreation were the main categories perceived as declining over time (18), but the sequence of restrictions occurrence was not investigated. More recently, transversal qualitative studies had similar findings from the perspective of adults with DM1 and their next-of-kin (19). In addition, the adults reported giving up more demanding activities (20). A deeper understanding of perceived changes and the sequence of occurrence of participation restrictions is needed, including what affects these changes. Quantitative and qualitative studies in DM1 have identified potential associations between participation restrictions and personal and environmental facilitators (21) and especially barriers (6, 21-31). Using technical aids and adjusting expectations regarding participation were the main facilitators (21). Muscle weakness and fatigue were among the primary barriers to participation (18, 21, 22, 24-29). Other barriers frequently found were: pain (22, 24, 25), limited family income (21, 24, 29), and lack of initiative or motivation (18, 20, 21, 30) for personal factors, as well as a lack of social support (18, 21, 24), perceived obstacles in the physical environment and accessibility (29) for environmental factors. However, most studies did not use a holistic conceptual framework to investigate these factors and did not capture an in-depth comprehensive picture of the processes underlying facilitators and barriers to changes in participation. Moreover, although multiple perspectives are required to understand a complex phenomenon like participation, most of these studies only described potential associations from the perspective of adults with DM1; a few (19,

20) added a relative's perspective, and none considered health professionals' views. To identify personal and environmental factors that act as facilitators and barriers to participation and explain how these factors interact, a better understanding of changes in participation and the underlying processes is needed from the perspective of the people directly involved. This study thus aimed to explore and explain changes in participation between diagnosis and the present time (15-37 years) from the perspectives of the adults with DM1, their relatives, and nurse case managers.

## **2. MATERIALS AND METHODS**

### ***2.1 Design and participants***

This study is part of a larger interdisciplinary longitudinal study characterizing DM1's natural progression over nine years (two measurements taken during this 9-year period), including a description of changes in participation and identification of predictors of change (7, 24, 29, 32). Structured by the Human Development Model – Disability Creation Process (HDM-DCP) framework, a qualitative multiple case study design (33) provided an in-depth description of each case, i.e., individual changes in participation and the involvement of personal and environmental facilitators and barriers. A purposive sample of six cases was recruited, with each informed by a triad: 1) an adult with DM1 (n=6), 2) a close relative (n=6), and 3) their nurse case manager from the Neuromuscular Clinic (n=3). The adults with DM1 were participants in the longitudinal study (7, 29). To be eligible for the current study, they had to: 1) have DM1 confirmed by genetic analysis with the juvenile or adult phenotype; 2) be 18 years of age or older; 3) speak French, 4) have a clinically significant change (improvement or decline of at least 0.5/9 (34) on the Assessment of Life Habits 3.1 [LIFE-H 3.1]; (35)) in three or more categories of



participation (12 categories in all) in the longitudinal study, and 5) have a close relative (mainly but not exclusively family members) and a nurse case manager who agreed to participate in the study. Since they had different prognoses, individuals with the other phenotypes of DM1 were excluded as well as those with another disease affecting participation (like stroke). Of the 115 participants from the longitudinal study, 87 were still alive at the time of this study and 49 met the inclusion criteria. A maximum variation sampling strategy (36) considering expert triage (37) was based on sex, age, and direction of participation change of the adults with DM1 (improvement or decline in participation). They were first contacted by a research assistant, invited to identify a significant relative (someone seen more than once a month for over ten years, including family and friends), and asked if their nurse case manager could be interviewed regarding her perception of their changes in participation over time. Prior to data collection, individual written informed consent to participate in the study was given by all participants. Grocery gift cards were given to the adults with DM1 and their relatives to compensate for their time. Nurse case managers were paid to participate by the Neuromuscular Clinic. The Ethics Review Board of the Saguenay–Lac-St-Jean Integrated University Health and Social Services Center approved the study protocol (#2015-006).

## ***2.2 Research context***

The study was conducted in Saguenay–Lac-St-Jean (eastern part of the province of Quebec, Canada) which has the highest prevalence (<158/100,000) of people with DM1 (38). Post-diagnosis, all study participants received health services from an interdisciplinary team coordinated by a nurse case manager from the local Neuromuscular Clinic. Quebec's healthcare system provides free province-wide health insurance. Like Quebec's other rehabilitation centers, the Neuromuscular Clinic uses the HDM-DCP as a main framework. This holistic framework

provides a conceptualization and operational definition of participation and considers interactions between personal and environmental factors that may facilitate or impede participation (8). For example, a restriction in participation arises when the social and physical environment cannot compensate for a personal impairment. The HDM-DCP also provides a nomenclature and classification for personal and environmental factors. The research group is located on the same floor as the Neuromuscular Clinic, but only one research team member (CG) has her office there. In the present study, the principal investigator (KR) was a doctoral candidate supervised by two senior professors/researchers with expertise in neuromuscular disorders and participation. All researchers were occupational therapists.

## ***2.3 Data collection***

### *2.3.1 Questionnaires*

Sociodemographic characteristics of all participants were collected using a generic questionnaire or extracted from their medical files. For adults with DM1, recalled time since diagnosis, self-rated health, ambulatory status, and participation level were also collected. To assess self-rated health, they answered the question: “How is your health generally right now?” on a scale of excellent, good, fair, or poor. Based on the HDM-DCP, participation was assessed using the LIFE-H 3.1 (35) for three measurement points: the first two extracted from the longitudinal study in 2002-04 (T1) and 2011-13 (T2) and the third collected at the end of data collection for adults with DM1 by the principal investigator in 2017-18 (T3). The principal investigator was trained to administer the LIFE-H by the occupational therapist who conducted the longitudinal study and, to limit interrater discrepancies, to use the same procedure and examples. The LIFE-H 3.1 is a 77-item questionnaire assessing level of accomplishment and satisfaction globally and for two domains, daily and social activities, each encompassing six categories. Daily activities categories

are nutrition, fitness, personal care, communication, housing, and mobility, while social activities encompass responsibilities, interpersonal relationships, community life, education, employment, and recreation. Since none of the adults with DM1 were at school or employed during the longitudinal or current study, the education and employment categories were not applicable. Accomplishment and satisfaction levels were assessed respectively with 10- and 5-point scales, where higher scores indicate greater participation and satisfaction. Used as one of the inclusion criteria for this study, a minimal clinically important difference (MCID) of 0.5 was previously set for accomplishment level (34). The LIFE-H presents excellent intrarater (intraclass correlation coefficients [ICC]: 0.80 to 0.91) and interrater (ICC: 0.86 to 0.92) reliability in the DM1 population for all scores except two because of very homogeneous scores (fitness: ICC of 0.20 and 0.21; communication: ICC of 0.12 and 0.47 (39)).

### *2.3.2 Semi-structured interviews*

The principal investigator conducted individual semi-structured interviews with each triad participant separately, at their home for adults and relatives and at the Neuromuscular Clinic for nurses. Adults with DM1 were interviewed twice, before and after other participants in their triad; each interview lasted about 60-90 minutes. Most relatives and nurses were interviewed once for about 60-90 minutes and 45-60 minutes, respectively. This interview sequence allowed the principal investigator to add specific questions in the second interviews with adults with DM1 based on information given by their relatives and nurses. All interviews were conducted with semi-structured interview guides developed for this study and adapted for each type of participant (adults with DM1, relatives, and nurses). To get a holistic understanding of participation, the interview guides were structured based on the HDM-DCP. Guides for relatives and nurses focused on their perspective of the adult with DM1's changes in participation. To

explore global changes in participation as well as the main facilitators and barriers and their interactions, the guide for the first interview with adults with DM1 included open-ended questions such as: “Since you were diagnosed with DM1, what changes have occurred in how you do your usual activities?” and “What does have caused these changes?” For the second interview, changes in specific activities as well as more specific facilitators and barriers were explored, with open-ended questions such as: “Describe to me how [*this activity*] changed” and “What does have caused the changes in [*this activity*]?” Before data collection, interview guides were reviewed by two DM1 research team members not involved in this study (MLa, a nursing professor/researcher, and MT, a doctoral candidate in social work). The interview guide for adults with DM1 was pretested with a patient/partner of the research group, resulting in minor changes to questions. The guides evolved during data collection and specific open-ended questions were added, such as “What is your motivation to do the activity now and before your diagnosis?”. Interviews were digitally audio-recorded and transcribed in full. Following the interviews, a synthesis of the discussion was mailed to each participant. A week later, the principal investigator followed up by phone to verify synthesis accuracy, ask further questions or make clarifications and corrections, if necessary. Only one change was made about the purchase of a technical aid in the synthesis of one adult with DM1.

### *2.3.3 Medical chart reviews*

Between the two interviews with adults with DM1, the principal investigator reviewed their medical charts from the Neuromuscular Clinic. An extraction grid (see Supplemental Appendix) was used to identify information about the sequence of occurrence of participation restrictions along with personal and environmental factors acting as facilitators or barriers. This information was retrieved from the diagnosis in the chart review and extracted from the clinical notes of the

interdisciplinary team (nurse, neurologist, physician, physical therapist, and occupational therapist).

#### *2.3.4 Artifacts*

Artifacts of important facilitators and barriers to participation identified in the environment of adults with DM1 were photographed by the principal investigator immediately after the first interview with them. Although they had consented in writing, participants also had to consent verbally before the artifacts were photographed and anonymized.

#### *2.3.5 Field notes*

Field notes, mainly audiotaped and handwritten, were used during data collection and analysis to document observations (e.g., interview context) and analytical thoughts about the cases.

### **2.4 Data analysis**

To explore and explain changes in participation along with the processes underlying facilitators and barriers in personal and environmental factors, a thematic content analysis (40) was performed using all the interview transcripts, medical charts, clinical interpretation of LIFE-H scores, artifacts, and field notes. An evolving coding manual and data extraction grids were used. Between the diagnosis and this study, changes in participation were identified qualitatively when an increase or decrease in level of accomplishment or satisfaction was reported by participants during the interview or documented in the medical chart. Changes in participation between the longitudinal study and current study were identified quantitatively when changes in LIFE-H scores were above the MCID, indicating an increase or decrease in accomplishment level. As the LIFE-H measures partly covered the current investigation's timeframe, they were used to enrich our overall understanding of changes in participation from the longitudinal study and to supplement information regarding the sequence of occurrence of participation restrictions.

Analysis and interpretation were monitored constantly to ensure rigor (41). In particular, codes were reviewed and discussed with the last two authors and another member of the research team (MT). Themes that emerged from the data were organized and renamed according to the HDM-DCP framework. The data saturation point appears to have been reached, i.e., no new themes emerged when the last case was added. To contextualize and compare the perspectives of each triad participant, an intra-case analysis of the interview transcripts, LIFE-H scores, medical chart reviews, artifacts, and field notes was first performed with pattern matching logic (42). As suggested by Yin (2014) (42), to identify recurring and contrasting themes from one case to the next, an intercase analysis was then carried out with cross-case synthesis logic, i.e., themes related to changes in participation and facilitators and barriers were compared using syntheses and matrices. Finally, to illustrate a temporal sequence of occurrence of participation restrictions, medical chart data were crossed in a chronological sequence with LIFE-H scores and interview transcripts if participants provided sufficient temporal markers (e.g., “I stopped dancing when I was 30-35”). QRS NVivo (version 12.0 for Windows; QRS International) was used for the analyses, Microsoft Word for the additional syntheses and matrices, and SPSS (version 25.0 for Windows; SPSS Inc) for the temporal sequencing.

### **3. RESULTS**

#### ***3.1 Demographics***

Of the eight adults with DM1 invited to participate, six accepted and two refused (not interested). Three women and three men between the ages of 40 and 56 participated in the study; the majority (4; 66.7%) had the juvenile form of DM1 (Table 1). All had a high school education or less and a variety of mobility capacities (from ambulant to wheelchair-bound); half rated their

health as fair. They presented multiple comorbidities and antecedents, the most common being musculoskeletal disorders, depression, anxiety, vascular disorders, and diabetes. The majority (4; 66.7%) of relatives were female family members, and half were over age 60. Contact frequency in the previous year between adults and relatives varied from at least once a day to twice a month. Nurse case managers were all women between the ages of 35 and 45. They had done in-person annual follow-ups with the adults with DM1 for approximately ten years and, when necessary, additional phone contacts.

[Please insert Table 1 about here]

### ***3.2 Changes in participation since DM1 diagnosis***

Six themes described the changes in participation between DM1 diagnosis and the present time. General changes included heterogeneous changes and an insidious increase in restrictions while specific changes encompassed redesigning accomplishment, progressive social isolation, restrictions in life-space mobility, and increasingly sedentary activities (Table 2). Each theme included up to three secondary themes.

[Please insert Table 2 about here]

#### ***3.2.1 General changes***

##### ***3.2.1.1 Heterogeneous changes***

Since diagnosis, all cases presented heterogeneous changes mainly characterized by a slow progressive decline in participation (Table 2) with varying degrees of restrictions depending on activities. LIFE-H results showed a clinically significant decline in domains and many categories for all adults with DM1 (Table 3). Between T1 and T2 (nine-year interval), as an inclusion criterion, they had to present a clinically significant change in at least three categories, but results showed various patterns of change with different categories affected in each case (see underlined

scores in Table 3). Between T2 and T3 (four-year interval), a possible acceleration in the decline was observed overall (see bolded scores in Table 3). Participation varied greatly from one category to another, leading to a heterogeneous portrait of changes over time, i.e., pattern of decline, improvement, stability, and a combination thereof.

[Please insert Table 3 about here]

**Improvements in participation occurred periodically** for five cases when there was something new in the environment (Table 2). For three cases, meeting new people resulted in more opportunities to do new activities. A5, who had a new boyfriend and new friends following a change in her living environment, reported having started going out again downtown and to restaurants and doing more recreational activities.

Due to fluctuations in some symptoms, **day-to-day variations in restrictions** in participation occurred for five cases. Daily fluctuations in capacity prevented planning trips or activities in advance, as explained by A2:

*“I don’t know if I’ll be able to go to my nephew’s birthday party. I [only] know from day to day. I can’t plan to do [activities], make future plans.”*

When comparing all cases, a **trend towards a temporal sequence** of occurrence of participation restrictions was identified (Figure 1). Three categories, i.e., education, employment, and responsibilities, tended to be restricted earlier in the disease course (about five years post-onset of the first symptoms). Personal care, community life, and communication tended to be restricted later (about 20 years after symptoms onset). For most categories, occurrence of participation restrictions after onset of the first symptoms varied greatly, from 5 to 25 years.

[Please insert Figure 1 about here]

### *3.2.1.2 Insidious increase in restrictions*



With the slow progression of disease, restrictions in activities occurred insidiously, i.e., so slowly they were often imperceptible in the person's daily life (Table 2). Most adults with DM1 described **feeling they had reached turning points** in restrictions in their participation over time. They suddenly realized they had difficulty doing a routine activity they previously considered possible, which enabled them to judge the real gravity of their situation and triggered recurring grief for the loss of their ability to do some activities. These activities were mainly related to nutrition, personal care, housing and infrequent recreational activities, which A5 illustrated as follows:

*“Two years ago, I used to make macaroni. [...] I thought I could [stir the macaroni], but when I picked up the spoon, I found that, without warning, I couldn't do it anymore.”*

A1 and Nb noticed that **restrictions were incorporated as if they were normal**. For the former, all activities were now done mechanically as he had progressively adapted to his condition and environment when the restrictions occurred. As for the nurse, she explained that, compared with many other adults with DM1, some people who fell often hardly noticed their falls since they happened every day. This insidious process was not recognized by A3, whose condition had deteriorated rapidly following multiple hospitalizations for respiratory infections.

### *3.2.2 Specific changes*

#### *3.2.2.1 Redesigning accomplishment*

All triads participant recognized the need and importance to rethink how to do activities based on individual interests and changes in abilities and environment (Table 2). Redesigning accomplishment involved adults with DM1 and also their relatives, depending on which activities they participated in. Except for two relatives, all participants noted that the six adults with DM1 had to **be more selective from an increasingly limited choice of activities** which

they wanted to do. More specifically, they had to do shorter or less demanding activities more often (such as preparing simple meals, looking after their pet, maintaining family relationships).

Activities considered less demanding varied with the person with DM1.

Adults with DM1 redesigned accomplishment by **modifying their activities**, such as by taking on a different role (e.g., becoming an advisor) or customizing activities so they could do them (e.g., breaking them up). For example, because of diminished ability to get up from a squat and carry things, A4 went grocery shopping more often, purchasing fewer items each time.

During the redesigning accomplishments process, **modifications in the environment** occurred gradually for nearly all cases. Depending on varying contributions from the main people concerned, these changes involved environmental adaptations, technical aids or human assistance (from relatives or community services). As time went by, progression in the amount of assistance required was noted (Figure 1). For many categories, especially nutrition and mobility, first-reported difficulties occurred before technical aids and human assistance were needed.

Environmental modifications were customized and varied from case to case, even for similar problems. For example, A2 and A4 were ambulant, but had similar difficulties with walking and going up and down stairs; the woman, who lived on the 2nd floor, had the stairs made non-slip, started to use a walking stick and wanted to buy ankle protectors, while the man happily moved to a single-storey apartment and used tibial braces and a cane.

#### *3.2.2.2 Progressive social isolation*

As time went by, social networks of adults with DM1 got smaller, leading to progressive isolation (Table 2). For some, current social networks were limited to one or three family members and one or two friends at most, as explained for A4 by Nb-4:

*“He is often lonely. [...] His network is his family. People with motor problems go out less, fewer people visit them, they self-isolate.”*

Nearly all cases had **lost friends over time** because of the disease, as reported by A6:

*“I don’t have friends any longer. They all [left] when they saw that I couldn’t do anything anymore.”*

After their network disintegrated, it was hard for nearly all cases to re-create, then maintain, a large social network.

Contributing to progressively socially isolated situations, all cases had **missed opportunities to maintain a social network**. Some talked about not being able to do social activities that typify adult life, like having children or being employed. A1 and A2 described the emptiness resulting from not having children because of the risk of disease transmission. All adults with DM1, except A5, reported feeling that they had missed employment opportunities having had a disability at work early in their lives. A4 and A5 also said they had missed recreational opportunities at various times (e.g., travelling with friends), which triggered feelings of loneliness and loss of meaning in life.

### *3.2.2.3 Restrictions in life-space mobility*

Over time, all cases exhibited more difficulty with going out often or going far from home (Table 2). First, all cases **went out less and less** regardless of the season, staying at home more often. Most considered staying at home a comfortable, easy alternative.

Second, four adults with DM1 **stayed closer to home**, raising issues of mobility and their environment impeding their ability to go out, as related by A1:

*“Before, I used to go to restaurants downtown, I’d take the bus [...] but now I go to the [fast food restaurant near me]. [...] It’s my monthly outing.”*

#### 3.2.2.4 Increasingly sedentary activities

Over time, activities became more sedentary, i.e., increase in activities done mostly sitting or lying down and requiring less energy. For five cases, watching television became the main activity in their occupational routine. Nb-3 described this for A3:

*“He watches TV. [...] He’s not as active as before. With the wheelchair and his weakness, it’s challenging to go out [and] do recreational activities.”*

While adults with DM1 do a selection of activities that are less demanding physically when redesigning accomplishments, are increasingly socially isolated and experience more and more restrictions in their life-space mobility, more sedentary behaviors occurred across all three previous themes.

### 3.3 Facilitators and barriers to participation over time and their underlying processes

Only slightly more than one quarter (11/37 factors; 29%) of personal factors were considered facilitators or both facilitators and barriers (Table 4). Conversely, nearly three quarters (18/25 factors; 72%) of environmental factors were facilitators or both. Processes underlying each facilitator and barrier accounted for participation changes (Table 4). They were also related to themes in the previous section 3.2 (Table 2) or to participation domains and categories (Table 3). However, interactions between facilitators and barriers in personal and environmental factors presented a more comprehensive explanation of participation changes through two distinct processes, namely the fit between factors and a potentially syndemic interaction.

[Please insert Table 4 about here]

In personal factors, the most important **facilitators** were related to the capacity for resilience (behaviors) of adults with DM1 and to how meaningful the activity was. To foster participation of adults with DM1 over time, support from family, adaptation and arrangement of the living

environment (home and community), and use of technical aids turned out to be vital for environmental factors (Table 4). R6 explained the importance of resilience for A6:

*“It took a long time before she accepted the disease. But now, she is able to overcome daily problems; find solutions when she encounters difficulty [in her activities]”*

The most common **barriers** in personal factors were related to greater perceived muscle weakness and fatigue varying from day to day. Effectiveness in redesigning accomplishment was affected by executive functions, especially problem-solving ability, which were important in modulating how activities were modified. In environmental factors, precarious social networks with a limited number of caregivers resulted in less assistance and fewer participation opportunities for adults with DM1. Participation was also restricted by financial abuse and domestic violence, which mostly affected women and were related to precarious social networks and the risk of being stigmatized by people in the community.

In addition, **participation** in an activity over time was influenced by how adults with DM1 viewed their adaptability. For example, activities considered easier to adapt continued to be done longer as the disease evolved.

The **processes** underlying facilitators and barriers were complicated by, on the one hand, the fit between facilitators and barriers in personal factors and environmental factors and, on the other, the presence of a potentially syndemic interaction. The **fit between facilitators and barriers**, both personal and environmental, with the activity modulated the impact of the symptoms on the participation of adults with DM1. For example, because he feared falling when using a wheelchair in a suboptimal environment (limited space with a floor-to-ceiling pole next to the toilet and bathtub; Figure 2), A3 decided to restrict his participation in bathing. He made this decision despite various facilitators, including (limited) ability to do sit-to-stand transfers,

presence of technical aids, and the availability of bathing assistance from the front-line healthcare center. Juggling with the fit between personal and environmental factors had induced him to start taking a sponge bath sitting in the kitchen a few months earlier.

[Please insert Figure 2 about here]

Interactions between personal and environmental factors through a **potential syndemic interaction**, i.e., co-occurrence of health and social conditions which worsen the condition of the person (43), was a barrier for all cases. DM1 symptoms associated with comorbidity (e.g., potential interactions at the cellular level) and barriers in the sociocultural environment (low income, stigma, etc.) exacerbated participation restrictions. Na offered the following general explanation:

*“All the symptoms [of DM1 and comorbidities] are obstacles for them. [...] Their family members do not understand the disease [or] are not considerate about the symptoms. Not to say about the attitude of some people in the community. To get out of the house, they have to overcome so many [obstacles].”*

Temporal interaction linked to the fit and to a potential syndemic interaction between facilitators and barriers in personal and environmental factors could account for the strongest trends in participation changes (Figure 3), including their heterogeneity.

[Please insert Figure 3 about here]

#### **4. DISCUSSION**

This study used a holistic framework to explore changes in participation of adults with DM1 from the perspective of the people directly involved, and explain changes with potential facilitators and barriers along with their underlying processes. Since their diagnosis, i.e., over

several decades, adults with DM1 experienced a slow, heterogeneous, and insidious decline in their participation. They redesigned how they did some or most of their daily and social activities, but progressive social isolation, restrictions in life-space mobility, and sedentary behaviors increased. The lack of fit between multiple personal and environmental factors (similar to Lawton's ecological model of aging (44)) and the co-occurrence of comorbidities and sociocultural obstacles (similar to the syndemic concept of health (43)) exacerbated participation restrictions over time. These results have implications in terms of the decline in participation since DM1 diagnosis, multifactorial nature of participation changes, and clinical practice.

#### ***4.1 Decline in participation since DM1 diagnosis***

Mainly characterized by a slow decline in participation post-diagnosis, the heterogeneous changes found in this study are in line with previous studies on the evolution of participation in slowly progressive neuromuscular disorders, including DM1 (6, 7, 19). The sequence of occurrence of restrictions in participation was consistent with the work in DM1 of Nätterlund *et al.* (2000) (16) and Boström *et al.* (2005) (15) that identified an increase in restrictions in housing and mobility over five to ten years, and in personal care and recreation after ten years. Progressive social isolation related to loss of social network was also highlighted in a previous study (18). In addition, the redesigning of accomplishment by adults with DM1 and some of their relatives is consistent with the modification of activities in previous studies (18-21). The efficacy of redesigning accomplishment was partly dependent on executive functions (e.g., problem-solving skills) of adults with DM1, which was also highlighted in other studies by individuals with DM1 (45) and their caregivers (46). In the current study, adults with DM1 and their relatives had redesigned their participation to constantly adjust to small day-to-day challenges and cope with progressive social isolation and life-space mobility restrictions. The insidious

increase in restrictions, with adults with DM1 facing turning points that reactivated their grieving process, might be attributable to minor adjustments made over an extended period. *Photovoice* used with nine adults with DM1 described a similar constant grieving process with cyclical pattern of loss (21). For some adults with DM1 in the current study, participation restrictions appeared to be normalized. Since DM1 is a hereditary disorder with many members of the same family having the disease (5, 47, 48) and disease awareness is known to be diminished in this population (49), the normalization process might rely on referents or individual models in the social network of adults with DM1 (50). Normalization, however, could lead to the trivialization of important and possibly life-threatening episodes, such as falls or coughs caused by dysphagia. Boström *et al.* (2004) (18) found a similar insidious decline in participation and perception of restrictions as part of “normal” everyday life for people with various neuromuscular disorders, including DM1.

Insidious increases in restrictions along with more sedentary behaviors might lead to progressive deconditioning and involuntary muscle disuse, exacerbating progressive weakness over time.

Recent advances in knowledge about the benefits of exercise training for adults with DM1 showed the potential to reverse deconditioning and disuse phenomena (51, 52). More research is needed to clarify the impact of sedentary behaviors and exercise training on adults with DM1.

#### ***4.2 Multifactorial nature of occurrence of changes in participation***

Although rarely described in the literature, some facilitators that enhanced participation of adults with DM1 were found among personal factors; most, however, were environmental factors. As in the present study, resilience was previously found to be an important factor that contributed to participation in DM1 (21, 28) and to community belonging in older adults (53). Many barriers found in this study were discussed individually in previous research, but were not all covered in



one study. In line with the present results, Ladonna *et al.* (2015) (21) described premature aging affecting body image and interpersonal relationships as well as limited finances as restricting participation in recreation. A lack of understanding from relatives and the public (20, 21) and social stigmatization (28) faced by adults with DM1 were similar to the barriers identified in this study, which increased the risk of participation restrictions. Of note, the present study found that some adults with DM1 tended to socially self-isolate because of concern about being stigmatized. Nevertheless other factors might contribute to progressive social isolation, such as phobic anxiety and lower self-esteem for most severe form of the disease (54), but they were not identified in this study.

This study highlighted a decline in participation over time influenced by personal and environmental factors. It is important to consider this when planning longitudinal studies or clinical trials exploring intervention efficacy. In fact, controlling for specific personal and environmental factors, especially comorbidities, living arrangement (adapted or not), use of technical aids, and social network size and quality, could be important when assessing outcomes in such studies.

Occurrence of participation changes was found to be multifactorial for adults with DM1, including via a potential syndemic interaction. Initially developed for transmissible diseases, the syndemic framework is increasingly used to understand complex non-communicable diseases, such as inflammatory arthritis (55) or type II diabetes (56, 57). Potential recognition that medical and environmental problems cluster and interact in the DM1 population could lead to improved care. For example, it would increase focus on: 1) enhancing health professionals' health promotion and disease prevention roles; 2) supporting a holistic nurse-led management of DM1 (58); 3) adding allied healthcare professionals (e.g., special educators, kinesiologist,

zootherapist) to interdisciplinary teams; or 4) adding macro-level interventions, such as community-based rehabilitation approaches that enable health professionals and stakeholders to build consistent interventions across health, education, livelihood, and social systems (59). Further research examining the syndemic framework in the DM1 population is needed before recognizing a formal syndemic interaction.

### ***4.3 Clinical practice implications***

The results highlighted potential ways to optimize participation over time by refining the evaluation process and maximizing interventions by capitalizing on facilitators and reducing barriers. This study found a progression in participation restrictions for the same participation category (i.e., difficulty first, then need for technical aids, then need for human assistance).

These results might help health professionals to **refine the evaluation process** and guide individuals with DM1 through upcoming stages in participation restrictions using a prognostic approach. As some of the participants in this study encountered delays in healthcare interventions, recognizing the progression of participation restrictions might help to implement a more timely approach, such as planning home adaptations or relocation. Of particular concern are financial abuse and domestic violence linked to precarious social networks, which should be explored more systematically, particularly for women, by health professionals to prevent abuse and participation restrictions. This was already highlighted for childhood phenotype in a retrospective study of Gagnon *et al.* (2017) (60), where risk for (sexual) abuse and compromised financial responsibilities were reported.

With redesigning accomplishment and the insidious increase in restrictions over time, adults with DM1 could benefit from health professionals' support to ease the burden of making constant readjustments and grieving. For example, **capitalizing on facilitators**, health professionals could

focus on resilience, problem-solving skills, and tips for staying alert to important episodes (e.g., falling or coughing) that could be trivialized afterwards. As restrictions appeared, seeking for care become important for their caregivers (61). Thus, an approach to inform and accompany them could also be proposed.

As stigma appears to be a barrier to participation, **reducing this barrier** by enhancing inclusion of people with DM1 in the community might be one way to sustain participation. Community education combined with social interactions at the individual level are often effective in countering stigma and fostering the social inclusion of various populations (62-64). In particular, taking action to increase stakeholders' and the public's awareness should include revisiting stereotypes of people with disabilities (63). Considering the precarious social networks in DM1, social interactions (62, 64) might also be used to increase opportunities for people with and without disabilities to participate in community or recreational activities.

#### ***4.4 Strengths and limitations***

To our knowledge, this is the first study to conduct an in-depth qualitative exploration with a holistic framework of changes in participation of adults with DM1 from the perspective of triads. Using the HDM-DCP framework helped to structure the main facilitators and barriers to participation over time and provided a holistic understanding of processes involved in participation restrictions. Rigor was ensured through data triangulation, extensive data collection, intra- and inter-case analyses, and monitoring analysis and interpretation (65). Inclusion of only six cases based on a subsample from a longitudinal study, however, limited the transferability of the results. Using a maximum variation sampling strategy and describing the research context offset this limitation by ensuring diversity in the purposely selected cases. Adults with DM1 with shorter disease duration or without a relative or nurse case manager follow-up might be

underrepresented. Important facilitators and barriers to changes in participation pertaining to different profiles of adults with DM1 might have been missed. Although the interviews fostered a general understanding of changes in participation, this phenomenon was explored retrospectively, meaning that a potential recall bias existed. In view of potential cognitive deficits, during the interviews with adults with DM1, a contextualization strategy was used to enhance participation recall over time. Data triangulation, a plurality of methods, and apparently reaching a data saturation point limited this bias and increased the credibility and plausibility of the study.

## **5. CONCLUSION**

This study explored and explained short- and long-term changes in participation from the perspective of the people directly involved. A heterogeneous, slow and insidious decline in participation leading to redesigning accomplishment was found to be a core element in understanding the changes over time. This study also highlighted the multifactorial nature of participation changes, where a misfit between personal and environmental factors and potential syndemic interactions led to participation restrictions over time for adults with DM1 and their relatives. A better understanding of participation changes, as well as facilitators and barriers, supports the decision-making of adults with DM1, relatives and health professionals concerning disease management with a prognostic approach. These results open up new avenues for research on resilience, stigma, and syndemic interactions that exacerbate participation restrictions in adults with DM1. Further research is needed with a larger sample; it could also benefit from interviewing a younger cohort of adults with DM1 and including multiple follow-ups. Using more than one conceptual framework (e.g., syndemic framework or International Classification

of Functioning, Disability and Health) to compare and contrast different interpretations may help to get a better understanding of the phenomenon. Since the global COVID-19 pandemic might have exacerbated the participation restrictions of people with DM1 and impact their future, health professionals need to intervene to limit their social isolation. Focusing on delivering essential rehabilitation services that help to adapt participation, managing symptoms with telemedicine (66), and supporting the resilience process could be crucial strategies during and after the pandemic.

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## Table legends

Table 1: Characteristics of triad participants in each case

CASE	1	2	3	4	5	6
<i>Adult with DMI</i>	<i>A1</i>	<i>A2</i>	<i>A3</i>	<i>A4</i>	<i>A5</i>	<i>A6</i>
Age (y)	56	40	53	50	45	52
Gender	M	W	M	M	W	W
Phenotype	A	A	J	J	J	J
Recalled time since diagnosis (y)	34	15	39	24	23	24
Actual time since diagnosis* (y)	33	15	37	24	24	24
Disease duration* (y)	34	19	39	34	30	35
Year of diagnosis*	1984	2002	1980	1994	1994	1994
CTG repetitions*	400	300	700	1,300	700	300
Education* (y)	12	6	11	12	11	12
Mobility: Ambulant indoor/outdoor	Y/N	Y/Y	N/N	Y/Y	Y/N	N/N
Self-rated health	Fair	Poor	Fair	Good	Poor	Fair
Living alone	Y	Y	N	N	N	Y
Residential status	Tenant	Tenant	Tenant	Tenant	Private home	Tenant
Comorbidities and antecedents						
Anxiety		x			x	
Depression		x			x	x
Diabetes	x			x		
Epilepsy			x			
Fibromyalgia		x				
Musculoskeletal disorders (e.g., bursitis)	x	x	x		x	x

Overweight						x
Pseudotumor cerebri					x	
Vascular disorders (e.g., atherosclerosis)	x					x

<i>Relative</i>	<i>R1</i>	<i>R2</i>	<i>R3</i>	<i>R4</i>	<i>R5</i>	<i>R6</i>
Age (y)	60	34	56	82	63	66
Gender	M	W	W	W	W	M
Relationship	Neighbor	Cousin	Sister	Mother	Mother	Ex-partner
Relationship duration (y)	20	34	53	50	45	25
Reported contact freq./previous year	1/week	3/week	1/week	1/day	2/month	1/day
Have DM1-phenotype, if applicable	N	Y-A	Y-A	N	N	N

<i>Nurse case manager</i>	<i>Na-1</i>	<i>Na-2</i>	<i>Nb-3</i>	<i>Nb-4</i>	<i>Nc-5</i>	<i>Nc-6</i>
Age (y)	35-45		35-45		35-45	
Gender	W		W		W	
Time since first seen (y)	16	16	10	9	10	10
In-person follow-ups (#/y)	1	1	1	1	1	2
Reported phone contacts (#/y)	<1	5-6	1	<1	2-4	3-4

\* Extracted from medical files

Notes. M = man; W = woman; J = juvenile; A = adult; Y = yes (ambulant); N = no (wheelchair-bound).

Participants in each triad: Adult with DM1 (A#); Relative (R#); Nurse case manager identification-case number (N\$-#)

Table 2: Exploration of changes in participation since diagnosis

Principal and secondary themes	Participants in each triad
1. General changes	
1.1 Heterogeneous changes	<i>All participants (A1-6; R1-6; Na-c)</i>
1.1.1 Periodic improvements	<i>A1, A2, A4-6; R1, R2, R5, R6; Na-1-2, Nb-4, Nc-5-6</i>
1.1.2 Day-to-day variations in restrictions	<i>A1-4, A6; R2; Na-2</i>
1.1.3 Trend towards a temporal sequence	<i>All participants</i>
1.2 Insidious increase in restrictions	<i>A1, A2, A4, A5, A6; R6; Na-2, Nb</i>
1.2.1 Sudden realization of restrictions (reaching turning points)	<i>A2, A4, A5, A6; R6; Na-2</i>
1.2.2 Restrictions incorporated as “normal”	<i>A1; Nb</i>
2. Specific changes	
2.1 Redesigning accomplishment	<i>All participants</i>
2.1.1 Activities selected from an increasingly limited choice of activities	<i>A1-6; R1, R2, R4, R6; Na-1-2, Nb-3-4, Nc-5</i>
2.1.2 Activities modified	<i>A1-6; R2, R3, R5; Na-2, Nb-4, Nc-6</i>
2.1.3 Environmental modifications	<i>A1-3, A5, A6; R1, R3, R5, R6; Na-1 Nb-3-4, Nc-5-6</i>
2.2 Progressive social isolation	<i>A1-6; R1, R2, R3; Na-1-2, Nb-3-4, Nc-6</i>
2.2.1 Loss of friends over time	<i>A1, A2, A4, A5, A6; R2; Nc-6</i>
2.2.2 Missed opportunities to maintain a social network	<i>A1-6; Nb-4, Nc-6</i>
2.3 Restrictions in life-space mobility	<i>A1-6; R1-4; N1-6</i>
2.3.1 Going out less and less	<i>A1-6; R1-4, Na-1-2, Nb-3-4, Nc-6</i>
2.3.2 Staying closer to home	<i>A1, A3, A4, A6; R1, R2; Na-1, Nb-4, Nc-6</i>
2.4 Increasingly sedentary activities	<i>A1-4, A6; R1-4; Na-2, Nb-3-4</i>

Note. A# = Adult with DM1 with relevant triad #; R# = Relative with relevant triad #; N\$-# = Nurse case manager \$ identification with relevant triad #, where applicable.



Table 3: Participation scores over time in each case

CASE	1	2	3	4	5	6
LIFE-H scores (T1 <sup>†</sup> /T2 <sup>†</sup> /T3)						
Global participation	7.4/7.5/7.0	8.1/8.3/7.5	7.9/7.8/5.4	8.6/8.3/7.6	7.5/7.3/6.2	7.0/6.6/5.5
Daily activities	7.9/8.1/6.8	8.0/8.3/7.2	7.9/7.8/5.1	<u>8.8/8.3/7.6</u>	7.5/7.3/5.8	6.8/6.6/5.7
Nutrition	8.8/9.0/7.5	8.8/9.0/8.8	<u>8.3/9.0/3.3</u>	9.0/9.0/6.0	8.0/8.3/6.5	<u>7.0/6.0/6.0</u>
Fitness	<u>8.3/6.5/6.0</u>	<u>8.5/6.5/8.3</u>	<u>6.8/4.7/7.3</u>	<u>9.0/6.0/9.0</u>	<u>8.0/7.5/7.3</u>	<u>6.5/5.3/5.0</u>
Personal care	<u>8.0/8.6/7.8</u>	<u>7.8/8.9/8.4</u>	<u>8.9/8.3/5.4</u>	9.0/8.9/8.1	<u>8.1/6.3/5.9</u>	<u>8.0/6.3/7.3</u>
Communication	8.5/8.7/8.7	9.0/9.0/8.5	9.0/9.0/8.8	9.0/9.0/9.0	<u>7.9/9.0/8.9</u>	8.8/8.9/8.8
Housing	<u>6.8/7.4/5.8</u>	<u>6.5/6.8/5.3</u>	<u>7.4/6.5/3.4</u>	<u>8.5/7.6/7.0</u>	<u>5.9/7.3/5.1</u>	<u>5.3/5.4/3.9</u>
Mobility	<u>7.3/6.5/5.0</u>	<u>8.0/9.0/3.8</u>	<u>6.4/8.0/2.4</u>	8.4/8.4/6.4	<u>7.5/4.0/1.0</u>	<u>4.4/5.3/3.0</u>
Social activities	6.6/6.7/7.4	8.2/8.3/8.0	7.8/7.8/5.9	8.3/8.1/7.6	7.6/7.4/6.5	<u>7.2/6.6/5.3</u>
Responsibilities	<u>8.7/8.2/8.7</u>	<u>8.5/8.0/8.0</u>	9.0/9.0/8.3	9.0/9.0/9.0	9.0/9.0/9.0	<u>8.0/8.8/7.9</u>
Interpersonal relationships	7.2/7.2/8.3	9.0/9.0/9.0	9.0/9.0/9.0	7.2/7.2/7.2	9.0/9.0/9.0	<u>7.7/9.0/7.2</u>
Community life	<u>9.0/7.7/8.4</u>	<u>8.0/9.0/7.9</u>	8.3/8.0/5.7	9.0/8.7/9.0	<u>9.0/8.1/5.3</u>	<u>8.0/4.0/4.7</u>
Recreation	<u>2.3/2.8/4.2</u>	8.7/8.4/7.3	<u>5.5/4.8/0.4</u>	<u>9.0/7.2/5.0</u>	<u>4.0/2.4/2.7</u>	<u>3.0/4.3/1.6</u>

<sup>†</sup> Extracted from longitudinal study

Notes. Clinically significant changes ( $|\geq 0.5|/9$ ) in LIFE-H are shown as underlined scores for the longitudinal study (T1-T2) and bold scores for changes between T2 and T3; a higher score indicates greater participation.

Table 4: Personal and environmental factors and interactions affecting participation since DM1 diagnosis, structured using HDM-DCP framework

Factors	Processes underlying facilitators and barriers	Triad participants
<b>Personal factors</b>		
<i>Identity factors</i>		
<i>1. Sociodemographic, economic, and cultural characteristics</i>		
<i>1.1 Age; 1.2 Sex; 1.3 Gender; 1.4 Nationality; 1.5 Marital status; 1.6 Number of children; 1.7 Family role; 1.8 Sibling rank; 1.9 Education; 1.10 Occupation type; 1.11 Professional status</i>		<i>No theme</i>
<i>1.12 Type of income; 1.13 Amount of income</i>		
Reliance on social assistance	(+ / -) Social assistance was considered helpful but, with a limited budget, not enough to participate in activities, principally but not limited to recreation. Reliance on social assistance was linked to restrictions in education and employment.	<i>A1-6 R5 Nb-4, Nc-5-6</i>
	(+) Budgeting activities and furniture with family income was important for nutrition, housing, mobility, community life, and recreation.	<i>A2, A3, A4, A6 R2, R4, R6</i>
<i>1.14 Sociocultural status; 1.15 Residential modality; 1.16 Residential status; 1.17 Legal citizenship status; 1.18 Administrative language</i>		<i>No theme</i>
<i>2. Sociocultural identity</i>		
<i>2.1 Sociocultural identity; 2.2 Language used; 2.3 Sexual orientation</i>		<i>No theme</i>
<i>2.4 Physical, functional and psychological characteristics</i>		
Invisible disease becoming visible	(-) Physical appearance changed over time (e.g., baldness, myotonic facies) and reduced satisfaction in interpersonal relationships.	<i>A4 R2 Na</i>
	(-) Invisible symptoms (such as fatigue, pain, apathy) hindered interpersonal relationships characterized by a lack of understanding from relatives. Participants linked the disease becoming visible over time to insidious increases in restrictions, progressive social isolation and stigma.	<i>A1, A6 R1, R2, C5 Na-1</i>
<i>2.5 Diagnosis</i>		
CTG repeats expansion size	(-) DM1 symptoms were perceived as more severe with higher CTG repeats expansion size. Worse symptoms could generally entail greater	<i>A2 R2 Na-2, Nb-4</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
	participation restrictions. Five participants did not know their CTG repeats expansion size.	
<i>2.6 Life story</i>		
Multiple hospitalizations due to DM1 and comorbidities	( - ) Rapid and major decline in overall participation following multiple hospitalizations due to pneumonia for one adult with DM1. Work cessation because of multiple operations due to DM1 or comorbidities for another adult with DM1.	<i>A3 R3, R6 Nb-3</i>
<i>2.7 Life objectives; 2.8 Values</i>		
<i>2.9 Beliefs</i>		
Beliefs in fragments of misinformation about health and DM1	( - ) Used fragments of misinformation to make decisions about participation (e.g., because DM1 causes “irreversible nerve death”, there was no point participating in exercise programs).	<i>A1-6 R1-4, R6 Nb-3</i>
<b><i>Organic systems</i></b>		
<i>1. to 13. All systems</i>		
Premature aging	( - ) Looking older than one actually was affected interpersonal relationships.  ( - ) Having less capacity and more medical complications earlier in life restricted overall participation, depending on residual capacity and medical complications.	<i>A2, R2  Nb-3, Nc-5</i>
<i>14. Morphology</i>		
Change in body mass index	( + / - ) Increase or decrease in weight over time led respectively to more or less difficulty in mobility and community life. Weight increase in particular decreased satisfaction in interpersonal relationships.	<i>A2, A4, A6 R2, R3, R6 Na-2, Nb-3</i>
<b><i>Capabilities</i></b>		
<i>1. Intellectual capabilities</i>		
<i>1.1 Consciousness</i>		
Excessive sleepiness	( - ) Being sleepy during the day and getting up early was a daily challenge and resulted in a general loss of opportunities to participate in activities. Participants linked excessive sleepiness to increasingly sedentary activities.	<i>A1-6 R2, R4, R5 N1-6</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
Diminished concentration	( - ) Did not want to drive because of limited concentration, leading to a life-space mobility restriction.	<i>A2</i>
Awareness of own strengths and limitations	( + ) Knowing own strengths and limitations helped to go at own pace when carrying out daily and social activities and ask for help when needed.	<i>A1, A2, A4, A5, A6 R1, R4, R6 Na-1-2, Nc-5-6</i>
<i>1.2 Mnestic skills</i>		
Mild memory problems	( - ) Forgetting a personal care appointment was the main perceived impact of mild memory problems. Forgetting a technical aid (e.g., cane) in public increased feeling of stigma from not feeling legitimate, which affected satisfaction with community life.	<i>A1, A2, A4</i>
<i>1.3 Thinking</i>		
Executive functioning	( + / - ) Problem-solving skills and judgment helped or restricted the adult with DM1's capacity to carry out daily and social activities, find adaptations, and be satisfied with their activities. Participants linked executive functioning to the effectiveness of redesigning accomplishment and resilience.	<i>A1, A2, A3, A5, A6 R2, R4, R5, R6 Na-1-2, Nb-4, Nc-5-6</i>
<i>2. Language capabilities</i>		
<i>2.1 Verbal skills</i>		
Dysarthria management	( + ) Speech improved when talking slowly, easing communication with others.  ( - ) Relatives perceived some difficulty understanding the adult with DM1 because "fast" and "slurred" speech made it harder to communicate with them. Relatives were ashamed to keep asking the adult to repeat him/herself.	<i>R4  R3, R5 Na-1, Nb-3</i>
<i>2.2 Expression skills</i>		
<i>2.3 Comprehension skills</i>		
Difficulty understanding others' reasoning	( - ) Difficulty understanding others' reasoning led to barriers in interpersonal relationships.	<i>A6 R6</i>
<i>2.4 Metalinguistic</i>		
<i>3. Behavioral capabilities</i>		
<i>3.1 Volition</i>		

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
Highly meaningful activities	( + ) The more meaningful the daily and social activities were for the adults with DM1, the more willing they were to do them, even in a risky or difficult situation. Could be linked to heterogenous changes.	<i>A1-6 R1, R2, R4, R5 Na-2, Nc-5</i>
Apathy (described as: <i>being lazy; having no interests</i> )	( - ) Living with apathy (reduced goal-directed behavior) led to initiating less daily and social activities. Lack of interest in activities increased over time for one adult with DM1.	<i>A1, A2, A4, A6 R2, R3, R6 Na-2, Nb-4</i>
<b>3.2 Affectivity</b>		
Little sense of belonging	( - ) Little feeling of belonging to the community led to accepting loneliness and fewer community life and recreation opportunities. Could be linked to progressive social isolation.	<i>A2, A4, A6 Na-1, Nb-4</i>
<b>3.3 Behavior</b>		
Developing resilience	( + ) Despite difficulties, participants developed the ability to press forward, one day at a time, to keep their spirits up and continue to participate in daily and social activities in other ways. Resilience was more difficult to achieve closer to DM1 diagnosis. Participants linked good resilience to redesigning accomplishment.	<i>All participants (A1-6; R1-6; Na-c)</i>
Using social skills (e.g., humor)	( + ) Having “something to say”, being “easy to get on with”, and having a sense of humor facilitated interpersonal relationships.  ( - ) All the women reported having difficulty with managing their emotions, which affected their interpersonal relationships.	<i>A1-5 R1-6 N1-6  A2, A5, A6 R2, R6 Na-2</i>
Adherence to healthcare recommendations	( + / - ) More or less adherence to healthcare recommendations facilitated or restricted participation in nutrition, personal care, housing, mobility, interpersonal relationships, and recreation. Adherence was influenced by the expectations of the adults with DM1 and the timing of the recommendations (e.g., timing was better after adults with DM1 recognized a difficulty that impacted a highly significant activity).	<i>A2, A4, A5 R3, R6 Na-1-2, Nb-4, Nc-5</i>
Fear of falling	( - ) Fear of falling and getting hurt or being unable to get up alone restricted personal care (e.g., bathing), mobility, community life, and recreation. Being confident in using his new technical aids, one participant did not see any impact of his fear of	<i>A1-6 R2, R6</i>

Factors	Processes underlying facilitators and barriers	Triad participants
	falling on mobility (e.g., continued to go outside). Participants linked fear of falling to restrictions in life-space mobility.	
<i>4. Sense and perception capabilities</i>		
<i>4.1 Interoceptive functions; 4.2 Proprioceptive functions</i>		
<i>No theme</i>		
<i>4.3 Exteroceptive functions</i>		
Reduced vision	(-) Ptosis and cataracts hindered participation in communication, mobility (e.g., driving car), and recreation.	<i>A2, A3 Nb-3</i>
<i>5. Motor activity capabilities</i>		
<i>5.1 Reflexes; 5.2 Static posture</i>		
<i>No theme</i>		
<i>5.3 Voluntary movements</i>		
Progressive weakness	(-) More and more difficulty executing upper and lower body movements, particularly grip and foot movements, led to restrictions in overall participation, and particularly in nutrition, fitness, personal care, housing, mobility (e.g., increased fall risk), and recreation. Participants linked progressive weakness to redesigning accomplishment, restrictions in life-space mobility, and increasingly sedentary activities.	<i>All participants</i>
	(-) Long time to recover after a physical activity (e.g., shoveling, cleaning up, exercising) meant it took hours or a few days to recover and be willing to do another activity.	<i>A2, A4, A5 R2, R6</i>
Less balance	(-) Less balance over time increased risk of falls, and fear of falling led to restrictions in mobility, community life, and recreation. Participants linked less balance to restrictions in life-space mobility.	<i>A1-5 R2, R3, R6 Na-1, Nb-3, Nc-5</i>
Myotonia	(-) Myotonia was perceived less frequently over time and had more impact during the early years of the disease on carrying and manipulating moderate to heavy objects. Restrictions were noted particularly in nutrition (e.g., difficulty eating), communication (e.g., speech less fluent), housing (e.g., difficulty filling cat's litter box), and interpersonal relationships (e.g., shaking hands for too long).	<i>A1-6 R5 Na-2, Nc-5-6</i>
	(-) When walking and trying to maintain balance before falling, knees suddenly became stiff and increased risk of falls.	<i>A2</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
<i>5.4 Mobility</i>		
Walking more slowly and decreasing walking distance	( - ) Difficulty walking led to increased activity modification and dependence on technical aids or human assistance to continue participating in community life and recreation. Participants linked difficulty walking to increase in redesigning accomplishment and restrictions in life-space mobility.	<i>A1, A2, A4, A5 R1, R5 Na-2, Nb-4, Nc-5</i>
Progressive difficulty getting up and walking	( - ) When difficulty getting up and walking increased too much, participants became more dependent on wheelchairs, thus restricting fitness (e.g., sleeping position), personal care, and mobility.	<i>A3, A6 R6 Nb-3, Nc-6</i>
Ability to walk up and down stairs	( + ) Maintaining the ability to walk up and down stairs increased opportunities to participate in housing, community life, and recreation.	<i>A2</i>
<i>5.5 Manual capabilities</i>		
Decrease in manual dexterity	( - ) More difficulty manipulating objects in daily and social activities increased how often they were dropped and led to a decrease in satisfaction and participation, particularly in nutrition, personal care, housing, and recreation.	<i>A2, A3, A5, A6 R2, R5 Na-2, Nc-5</i>
<i>5.6 Praxes</i>		
<i>No theme</i>		
<i>6. Breathing capabilities</i>		
Shortness of breath	( - ) More difficulty and more time needed to do household chores in housing and to go out to participate in community life and recreation.	<i>A2, A3, A4, A5, A6 R2, R3 Na-1-2, Nb-3-4</i>
<i>7. Digestive capabilities</i>		
Appearance of dysphagia over time	( - ) More and more choking decreased satisfaction with nutrition.	<i>A1, A3, A4 R5 Nb-3-4, Nc-5</i>
Shift between constipation and diarrhea	( - ) Even after taking medication or modifying activities, shift between constipation and diarrhea hindered nutrition (e.g., food choices), interpersonal relationships (e.g., visiting family and friends), and community life (e.g., going to an appointment or shopping).	<i>A2, A4, A5 R2, R5 Na-1-2, Nb-4</i>
<i>8. Excretion capabilities</i>		
Fecal and urinary incontinence	( - ) Participants modified nutrition, community life and recreation activities to lessen the risk of	<i>A2, A5, A6 R2, R5, R6</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
	having incontinence in public. Acceptance of using protective pads depended on the attitude of the adult with DM1. A nurse suggested that her male patient might not talk about incontinence because of being embarrassed.	<i>Na-1</i>
<i>9. Reproductive capabilities</i>		
Sex drive	(+) Having an active sex drive increased satisfaction with sexual relationships.	<i>A2, A5 R2</i>
	(-) Loss of a partner's sex drive due to a shift in the relationship (e.g., feeling more pity) restricted sexual relationships.	<i>A6 R6</i>
<i>10. Protection and resistance capabilities</i>		
Day-to-day variations in level of physical fatigue	(-) Physical fatigue limited endurance for activities in all daily and social domains (especially employment), entailed breaking them down into less demanding parts, and increased the time needed to do them. Participants linked physical fatigue to redesigning accomplishment, restrictions in life-space mobility, and increasingly sedentary activities.	<i>All participants</i>
Day-to-day variations in level of pain	(-) Days with more pain increased irritability and participation restrictions in daily and social activities (various regions were painful, mostly back, knees, and shoulders).	<i>A1-6 R2, R3, R4, R6 Na-2, Nb-3, Nc-6</i>
Cold intolerance	(-) In winter, participants had more difficulty going outside in the community because of limited tolerance for cold. Cold intolerance was linked to restrictions in life-space mobility.	<i>A1, A2, A3, A5, A6 R2, R6</i>

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## **Environmental factors**

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### ***1. Social***

#### *1.1 Political/economic factors*

##### *1.1.1 Political system and government structures*

Difficulty advocating at the government level	(-) Desire for more opportunities and greater understanding from the government to facilitate overall participation with laxer eligibility criteria for social programs and greater financial aid.	<i>A3, A5 R5 Nc-5</i>
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##### *1.1.2 Legal system*

*No theme*

##### *1.1.3 Economic system*



<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
Perceived financial strain	( + ) Used strategies (e.g., bargaining, buying on sale) to ease financial strain with redesigning accomplishment.	<i>A2-6</i> <i>R6</i>
	( - ) Costs associated with nutrition (e.g., restaurants), mobility (e.g., taxi, bus) and for activities in the community (e.g., cinema) limited opportunities in community life and recreation.	<i>A1-5</i> <i>R2, R4</i> <i>Na-2</i>
Employment opportunities and flexibility	( +/- ) Using employment placement programs helped to learn job. Lack of understanding of employer created problems maintaining employment.	<i>A4</i> <i>R2, R4</i> <i>Na</i>
<i>1.1.4 Social/health system</i>		
Public or private adapted living environment needed	( + ) It was important to live at the “right” place and have a flexible living environment where services could be combined with disease progression to maintain satisfactory participation overall.	<i>A1, A3, A5</i> <i>Na-1, Nb-3, Nc-5</i>
	( - ) Lack of adapted living resources for people with a need for mainly physical assistance led to difficulty finding the “right” place to live.	<i>Nc-5</i>
DM1-related health promotion and prevention efforts	( + ) More information concerning DM1 has been given to frontline healthcare providers to facilitate disease monitoring and maintain overall ability to participate.	<i>Na</i>
	( - ) General public often not familiar with DM1 and lack of understanding hindered participation in interpersonal relationships and community life.	<i>A1, A2, A4</i> <i>R5</i> <i>Na</i>
Healthcare improvement over time	( + ) Having frequent and personalized follow-ups with a nurse case manager and/or family doctor facilitated navigation through the healthcare system and receiving services, which then optimized overall participation.	<i>A1-6</i> <i>R5, R6</i> <i>Na-1, Nb-3, Nc-5-6</i>
	( + ) Minimization of the complexity of procedures and documentation when interacting with frontline healthcare providers facilitated accomplishment of nutrition, personal care, and responsibilities. Help from nurse case manager was available if needed.	<i>A1</i> <i>Na, Nb-3</i>
Gaps in rehabilitation services	( - ) Lack of resources to promote activities with personalized support, such as a special educator to provide coaching on daily or social activities.	<i>Na</i>
Importance of social assistance	( + ) Local community service centers were crucial to support people and their caregivers (e.g., at-home	<i>A1-6</i> <i>R1, R3, R5</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
	support, respite services), coordinate services from community organizations (such as for household help or financial aid), and help people participate in personal care and housing.	<i>Na-1, Nb-3, Nc-6</i>
<i>1.1.5 Education system</i>		
Education issues	( - ) Intimidation and learning difficulties at high school led to dropping out.	<i>A2</i>
<i>1.1.6 Public infrastructures</i>		
Efforts to achieve universal accessibility with infrastructure adaptations	( + ) Increased urban planning and accessibility of local facilities (e.g., safer sidewalks, automatic doors) helped with accessing and navigating to participate in mobility, community life and recreation.	<i>A1 R6 Nb</i>
	( - ) Lack of consistency of some infrastructure adaptations (e.g., no snow removal on down slopes or sidewalks, no elevators in buildings with multiple floors, locked automatic doors, small spaces with no access for wheelchair users) led to restrictions in mobility and community life. Participants linked accessibility to restrictions in life-space mobility.	<i>A1, A6 R1 Nb</i>
Improvements needed in transportation services (public, private and adapted)	( + ) For mobility participation, alternatives were available when people stopped driving (e.g., lifts from relatives, public transportation).	<i>A1, A3, A5, A6 R1 Na-1</i>
	( - ) Multiple barriers were reported to the use of public, private, and adapted transportation (e.g., not all destinations possible for public and adapted transportation, high cost to use private services, need to plan to reserve adapted transportation services in advance, difficulty to use in winter conditions, rides often long and chance of incontinence (two adults with DM1 had bad personal experiences)).	<i>A1, A2, A3, A5 R3 Na, Nb-3</i>
<i>1.1.7 Community organizations</i>		
Importance of community organizations	( + ) Social and community organizations were crucial to provide participation opportunities and financial assistance programs (e.g., Meals on Wheels or technical aid programs from Muscular Dystrophy Canada).	<i>A1-6 R1, R3, R5 Na-1, Nb-4, Nc-5</i>
	( - ) Lack of knowledge concerning existing resources led to loss of opportunities to globally participate.	<i>A5</i>

Factors	Processes underlying facilitators and barriers	Triad participants
	(-) Delay before being eligible for community assistance programs (e.g., long waiting list or too many eligibility criteria) delayed participation in specific activities (e.g., nutrition, housing, mobility, recreation).	<i>A5, A6</i>
	(-) Lack of community resources in some regions reduced opportunities to receive help and restricted overall participation.	<i>A1, A3</i> <i>Na-2, Nb-4, Nc-6</i>
<i>1.2 Sociocultural factors</i>		
<i>1.2.1 Social network</i>		
Small, precarious social network	(+) Relatives had an essential role to help carry out activities, mainly in the daily activities domain (nutrition, personal care, housing, mobility), to motivate the adult to overcome a lack of interest, and to support psychologically. Non-judgmental attitude, proactivity, availability, and trust from the caregiver together with flexibility, striving for independence, and grateful attitude from the adult with DM1 facilitated the caregiving relationship.	<i>All participants</i>
	(+) Having complementary and alternate caregivers helped to choose the “right” caregivers for the right activities (good match is important to minimize the burden on caregivers).	<i>A4, A5, A6</i> <i>R4, R5, R6</i> <i>Nc-5</i>
	(+) Meeting new people enhanced participation in interpersonal relationships, community life and recreation, with new opportunities and an outside stimulus to participate. Participants linked meeting new people with periodic improvements.	<i>A5</i> <i>R1, R3, R5</i> <i>Nc-6</i>
	(-) Often limited to a few family members and friends, the social network was precarious. This meant few alternate caregivers (who are hard to replace) and less opportunities to participate in interpersonal relationships and community life. Participants linked the precariousness of the social network to progressive social isolation.	<i>All participants</i>
	(-) Greater burden on caregivers led to one couple’s separation and loss of opportunities to participate in interpersonal relationships.	<i>R6</i> <i>Nc-6</i>
	(-) All women with DM1 suffered from financial abuse and domestic violence, which affected how they participated in interpersonal relationships.	<i>A2, A6</i> <i>R5</i> <i>Nc-5</i>
Importance of pets	(+) Pets such as dogs and cats reduced loneliness.	<i>A1, A2, A3, A4, A6</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
		<i>R6</i> <i>Na-2</i>
<i>1.2.2 Social rules</i>		
Importance of values	( + / - ) In the circle of the adult with DM1 (person and relative), what is valued influenced how they participated globally (e.g., valued independence versus mutual aid).	<i>A2, A3, A5, A6</i> <i>R6</i> <i>Na-1, Nb-4, Nc-5-6</i>
Attitudes of people in the community (e.g., stigma)	( + ) Spontaneous help from people enabled wheelchair users to access community facilities or to get up after a fall in community life.	<i>A2, A4, A6</i> <i>R2, R5, R6</i>
	( - ) Being concerned about what people think or feeling stigmatized when being stared at decreased satisfaction with doing an activity in public or in a community setting, particularly when an assistive device labeled them as “disabled”. As two women said, they stayed at home to avoid people’s stares; attitudes of people in the community were linked to progressive social isolation.	<i>A2, A4, A5, A6</i>
<b>2. Physical</b>		
<i>2.1 Nature</i>		
<i>2.1.1 Physical geography</i>		
Avoiding hills and sloping streets	( - ) Activities in an area with hills and sloping streets were avoided over time due to difficulty walking. Participants linked avoiding hills and sloping streets to restrictions in life-space mobility.	<i>A1, A2, A6</i> <i>Nb-4</i>
<i>2.1.2 Climate</i>		
Winter	( - ) Winter led to mobility restrictions, fewer opportunities to participate in community life or recreation, higher risk of falls, less satisfaction because of intolerance to cold, more loneliness, and greater psychological distress. Participants linked winter conditions to heterogenous changes, progressive social isolation, and restrictions in life-space mobility.	<i>A1-6</i> <i>R1, R2, R5, R6</i> <i>Nb-4, Nc-5-6</i>
<i>2.1.3 Time</i>		
Delay in healthcare interventions	( - ) Delays in home relocation (took place two years after first request) led to multiple moves and impeded participation in personal care, housing, community life, and recreation. Delays in home adaptations (done five years after first request) led to	<i>A5, A6</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
	difficulty with personal care and multiple falls in the bathroom waiting to be adapted.	
<i>2.1.4 Sound; 2.1.5 Electricity and magnetism; 2.1.6 Lighting</i>		<i>No theme</i>
<i>2.2 Environmental development</i>		
<i>2.2.1 Architecture</i>		
Living arrangement	( + ) Living on the ground floor or in a residence with elevators facilitated mobility at home (in the housing category) when walking became difficult.	<i>A1, A3, A4, A5, A6 R2</i>
	( + / - ) Home adaptations and adjustments became mandatory at some point to participate safely in personal care and housing (e.g., less risk of falling at home). Participants linked a helpful living arrangement to periodic improvements.	<i>All participants</i>
Learning where to go in the community	( + ) Knowing which facilities and public buildings are adapted helps to choose the easiest place to go to when participating in community life. Participants linked learning where to go to redesigning accomplishment.	<i>A1, A5 Na-1</i>
<i>2.2.2 National and regional development</i>		
Proximity of facilities and relatives	( + ) Living close to facilities and relatives became more important over time, allowed more freedom and opportunities to do more activities in interpersonal relationships, community life, and recreation. Participants linked the proximity of facilities and relatives to periodic improvements.	<i>A1-4 R3-6 Nb-3-4. Nc-5</i>
<i>2.2.3 Technology</i>		
Driving cessation	( - ) Using a car less frequently combined with major cost to own and license a car led to voluntary driving cessation, which resulted in less participation in community life and recreation. Participants linked driving cessation to a restriction in life-space mobility.	<i>A1, A6 Nc-6</i>
	( - ) Disease progression led to driving evaluation and could result in driving cessation, which led to less participation in community life and recreation.	<i>A3, A4 R3</i>
Technical aids used	( + ) Willingness to use technical aids helped with nutrition, personal care, housing, mobility, community life, and recreation. Participants linked technical aids to periodic improvements, helping them to regain a capacity (such as improved walking with orthosis) and make redesigning	<i>All participants</i>

<b>Factors</b>	<b>Processes underlying facilitators and barriers</b>	<b>Triad participants</b>
	accomplishment. Technical aids had to be viewed as acceptable, useful, and cost-efficient by the adult with DM1.	
	( + ) Three- or four-wheeled scooter was both a recreational activity in the summer and a mobility facilitator linked to periodic improvements.	<i>A1, A3, A5, A6 R1, R6 Na-1, Nb-3, Nc-5-6</i>
	( - ) Some technical aids became obsolete or got broken, which restricted participation in specific activities.	<i>A1, A3, A5, A6 R3</i>
Potential of the technology	( + ) Use of technology (e.g., computer, video game, tablet, smartphone) was perceived as helping to maintain capabilities, facilitate responsibility for paying bills, and maintain relationships with others. Participants linked technology to periodic improvements.	<i>A1, A2, A4, A5, A6 R1 Nb-4</i>
<b>Interactions and temporal flux</b>		
Personal and environmental facilitators and barriers fit with the activity	( + / - ) Fit or lack of fit between multiple personal and environmental facilitators and barriers when doing an activity. A misfit between personal capacities and environmental demands with the activity led to a participation restriction.	<i>All participants</i>
Potential syndemic interaction	( - ) Comorbidities in co-occurrence with sociocultural environmental obstacles had the potential to worsen the condition of the adult with DM1 which could have adverse health consequences and exacerbate participation restrictions.	<i>A1-6 R2, R6 Na-1-2, Nb-3, Nc-5-6</i>
<b>Participation</b>		
Adaptability of activity	( + / - ) Some activities were viewed as harder (e.g., dancing) or easier to adapt (e.g., meal preparation, grocery shopping). Adaptability of activity influenced the redesigning of accomplishment over time.	<i>All participants</i>

Notes. A# = Adult with DM1 + relevant triad #; R# = Relative + relevant triad #; N\$-# = Nurse case manager \$ identification + relevant triad #, where applicable.

( + ) = facilitator; ( - ) = barrier.

Supplemental Appendix: Extraction grid for medical chart review

Participant case no \_\_\_\_\_

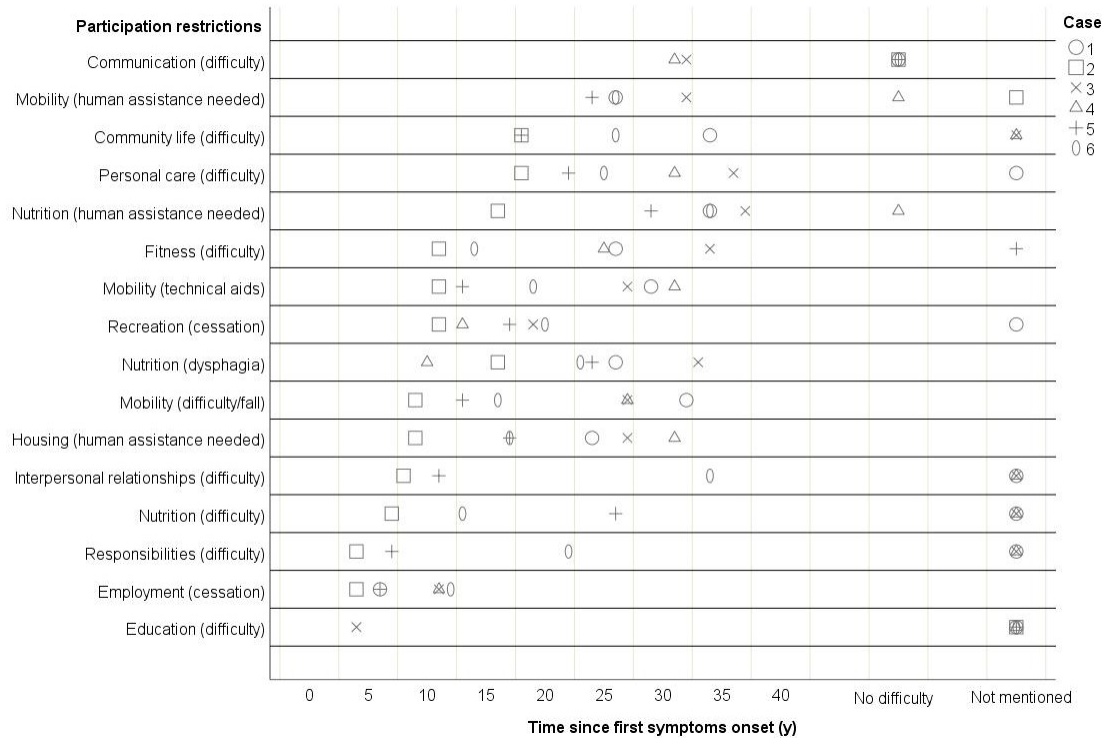
Date \_\_\_\_\_

Clinical notes of: nurse, neurologist, physician, physical therapy or occupational therapist (circle)

**TIMELINE** →

Date	Before (if reported)	Diagnosis	After				
Participation changes							
Personal factors	facilitators						
	barriers						
Environmental factors	facilitators						
	barriers						

## Figure captions



Figure

1: Temporal sequence of participation restrictions since first symptom onset. *Notes. Complete description of participation restrictions in order from bottom to top: Education (first-reported difficulty with education); Employment (first-reported employment cessation); Responsibilities (first-reported difficulty with budget or caring for family member); Nutrition (first-reported difficulty handling kitchen objects); Interpersonal relationships (first-reported difficulty with family relationships); Housing (first request for household assistance service or first-reported human assistance); Mobility (first-reported walking difficulty or fall); Nutrition (first-reported episode of dysphagia); Recreation (first-reported cessation of active or social recreational activity); Mobility (first request for mobility aids, not including electric wheelchair or 3- or 4-wheeled electric scooter); Fitness (first-reported episode of sleep disturbance); Nutrition (first request for meal delivery service or first-reported human assistance); Personal care (first-*



*reported difficulty with dressing or washing); Community life (first-reported difficulty with using a business (shopping) or participating in a community group); Mobility (first request for electric wheelchair or 3- or 4-wheeled electric scooter or adapted transportation service or first-reported human assistance); Communication (first-observed signs of dysarthria affecting communication).*



Figure 2: Suboptimal environment of case 3 when participating in bathing activity. *Note. Adult with DMI was unable to get close enough to the bathtub with his wheelchair to transfer safely because of limited space, combined with placement of toilet and bathtub, and a floor-to-ceiling pole.*

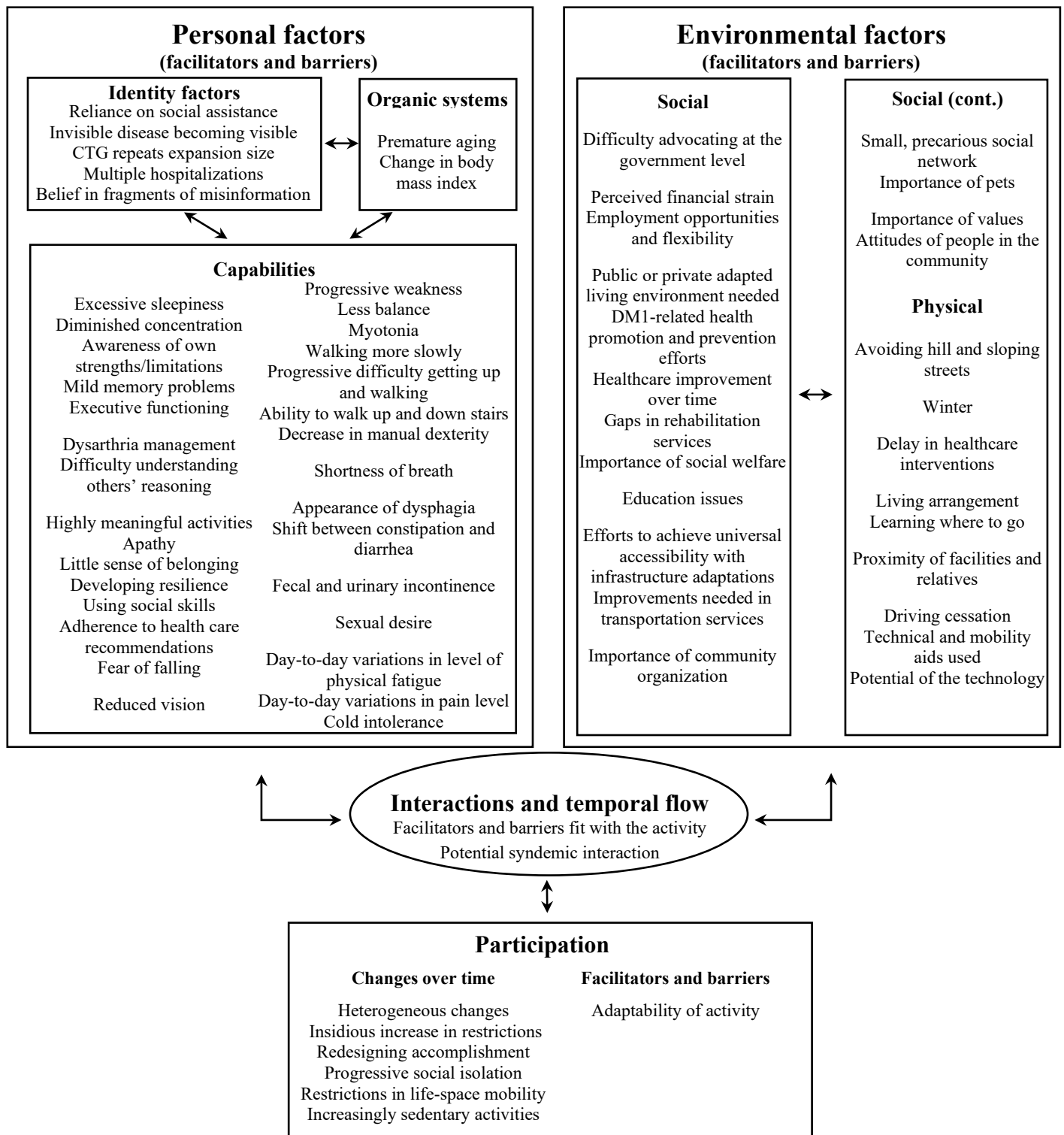


Figure 3: Synthesis of changes in participation over time, and facilitators and barriers for adults with DM1, based on adapted version of HDM-DCP framework.

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