

Efficacy of manual wheelchair skills training for improving the skills and confidence of people with hereditary degenerative disorders: Protocol of a sequential multi-methods approach

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Abstract

Background: Mobility impairment and participation restrictions are experienced by individuals with autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) and myotonic dystrophy type 1 (MD1), two disorders that are highly prevalent in the province of Quebec, Canada. People with ARSACS and MD1 experience a progressive decline in mobility, which commonly results in the provision of manual or power wheelchairs. While wheelchairs can facilitate mobility and social participation, provision alone does not guarantee safe and effective use. Wheelchair skills training has been shown effective for improving manual wheelchair (MWC) skills and confidence among adult MWC users with variable diagnoses, which may enhance self-directed mobility and participation and reduce the risk of chronic and acute injuries. However, MWC skills training for people with ARSACS and MD1 remains understudied.

Objective: The primary objective of this study is to evaluate the efficacy of MWC skills training for safely improving MWC skills performance in people with ARSACS and MD1. Secondary outcomes include exploration of the influence of MWC skills training on MWC skills capacity, MWC use self-efficacy, MWC mobility, and retention of outcomes three months later. We will also qualitatively explore the MWC training experiences of people with ARSACS and MD1.

Methods: This study will use a sequential multi-methods design, combining a waitlist randomized controlled trial and qualitative interviews. The participants will include adults who have a diagnosis of ARSACS or MD1, and who use a MWC for mobility. Participants will be randomly assigned to the intervention or control group using a 1:1 allocation ratio. The intervention group will receive 5 MWC skills training sessions (1-2 sessions/ week), while the control group will receive no training. Data will be collected at baseline (T1), after the 4-week intervention (or waiting period for the control group) (T2) and 3 months after T2 to assess retention (T3). The primary outcome will be MWC skills performance. Secondary outcomes will include MWC skills capacity, MWC use self-efficacy, and MWC mobility. Semi-structured individual interviews will be conducted to explore participants' expectations regarding MWC use, past MWC experiences and perceptions of MWC skills training. Quantitative data will be analysed using ANCOVA (controlling for baseline scores) and qualitative data using reflexive thematic analysis.

Results: Research ethics was approved in July 2024. Study teams members have met to plan recruitment which will start in Fall

2024. A graduate student, research assistant, and research coordinator have been recruited and trained.

Conclusions: MWC skills training may be one way to improve self-directed mobility and related outcomes for people with ARSACS and MD1. The results of this randomized wait-list controlled trial will help guide clinical practice toward MWC skills training for understanding potential influences on MWC mobility of people living with neuromuscular disorders. Clinical Trial: Clinicaltrials.gov NCT06596850

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Original Manuscript

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Abstract

Background: Mobility impairment and participation restrictions are experienced by individuals with autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) and myotonic dystrophy type 1 (MD1), two disorders that are highly prevalent in the province of Quebec, Canada. People with ARSACS and MD1 experience a progressive decline in mobility, which commonly results in the provision of manual or power wheelchairs. While wheelchairs can facilitate mobility and social participation, provision alone does not guarantee safe and effective use. Wheelchair skills training has been shown effective for improving manual wheelchair (MWC) skills and confidence among adult MWC users with variable diagnoses, which may enhance self-directed mobility and participation and reduce the risk of chronic and acute injuries. However, MWC skills training for people with ARSACS and MD1 remains understudied.

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Methods: This study will use a sequential multi-methods design, combining a waitlist randomized controlled trial and qualitative interviews. The participants will include adults who have a diagnosis of ARSACS or MD1, and who use a MWC for mobility. Participants will be randomly assigned to the intervention or control group using a 1:1 allocation ratio. The intervention group will receive 5 MWC skills training sessions (1-2 sessions/ week), while the control group will receive no training. Data will be collected at baseline (T1), after the 4-week intervention (or waiting period for the control group) (T2) and 3 months after T2 to assess retention T3). The primary outcome will be MWC skills performance. Secondary outcomes will include MWC skills capacity, MWC use self-efficacy, and

MWC mobility. Semi-structured individual interviews will be conducted to explore participants' expectations regarding MWC use, past MWC experiences and perceptions of MWC skills training. Quantitative data will be analysed using ANCOVA (controlling for baseline scores) and qualitative data using reflexive thematic analysis.

Results (Progress to date): Research ethics was approved in July 2024. Study teams members have met to plan recruitment which will start in Fall 2024. A graduate student, research assistant, and research coordinator have been recruited and trained.

Conclusions: The results of this randomized wait-list controlled trial will confirm whether MWC skills training may can improve self-directed mobility and related outcomes for people with ARSACS and MD1. Findings may help guide clinical practice toward MWC skills training for understanding potential influences on MWC mobility of people living with neuromuscular disorders.

Keywords: ARSACS; Myotonic Dystrophy Type 1; Manual Wheelchair; Wheelchair Skills Training; Mobility; Participation; Randomized Controlled Trial

Clinicaltrials.gov NCT06596850, 2024-09-19, Version 1

Introduction

Mobility impairment and participation restrictions are commonly experienced by people with neuromuscular and hereditary degenerative disorders [1]. Two highly prevalent disorders in the province of Quebec include Autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) [2,3], and Myotonic dystrophy type 1 (MD1) [4,5]. Decline in mobility is a hallmark among people with ARSACS and MD1, which can negatively influence physical and psychosocial health outcomes. Approximately 74% of individuals with MD1 have mobility impairments that restrict participation in activities of daily living, leisure activities, and vocation [6-8].

Wheelchairs are prescribed to facilitate mobility for people with ARSACS and MD1 when walking becomes difficult. In the reported literature, approximately 45% of individuals with ARSACS used wheelchairs (WC) [9-11], with the average age of WC provision at 38.9 years [10]. Given the varying rate of progression of ARSACS, wheelchairs may be suggested during adolescence or older age (ex. 17 to 59 years of age) [12], but most people (if not all) with eventually need a WC for mobility. A patient registry of MD1 in the United Kingdom reported 56% of patients used a wheelchair, with the average age at which wheelchair use began at 34.6 years, ranging from infancy to old age (1 to 72.4 years). Among the wheelchair users in this registry, 22.8% started using a wheelchair before the age of 10 years and 56.6% started between 30 and 59 years of age [13].

Regardless of diagnosis, WC provision alone does not guarantee safe and effective use. If not used adequately, WCs could be associated with mobility dependence, restricted participation, reduced social connectedness [14], and reduced quality of life [15]. Moreover, the unsafe use of WC could lead to accidents that vary in severity from cuts and bruises to life-threatening head injuries [16]. Furthermore, injuries due to WC accidents may increase caregiver burden and strain on the

healthcare system [17].

Learning how to use the WC and developing appropriate WC skills are critical to independent mobility and participation [18]. However, WC mobility for people with ARSACS and MD1 remains understudied. Our team recently demonstrated that adults with ARSACS had lower WC skills but fewer training goals than adults with other diagnoses (e.g., spinal cord injury) [19]. This disparity may be attributed to limited training opportunities and low expectations of WC use for individuals with ARSACS [19]. It may be that the progressive nature of ARSACS and MD1 (and other neuromuscular disorders) and the clinical focus on maintenance of functional walking has historically taken clinical precedence over WC mobility outcomes.

Training in how to use WCs and follow-up are suggested by the World Health Organization as important steps of WC service provision [20,21]. Four meta-analyses demonstrate the effectiveness of WC skills training for improving WC skills, WC use confidence and participation [21-24]. However, a survey of 68 Canadian rehabilitation centres demonstrated that > 50% of therapists spent two hours or less on WC skills training and 18% provided no training at all [25]. Moreover, individuals with ARSACS and MD1 do not typically spend periods of time in rehabilitation centres where most WC skills training occurs. The inherited and progressive nature of ARSACS and MD1 means that people receive WC when they can no longer walk, but they commonly do not receive additional assessment, training or follow-up related to WC use. Given evidence of the effect of WC skills training in other progressive conditions (eg, multiple sclerosis, Parkinsonism) [23,24], the association between WC mobility and the preservation of upper limb function and strength in ARSACS [19], it is likely that evidence-based training could improve WC skills, confidence and social participation for people with ARSACS, MD1 and other neuromuscular disorders. In a pilot study, 3 individuals with ARSACS improved their manual WC (MWC) skills (18-30%) and

confidence (5-15%) after 3 hours or less of MWC skills training [26].

The overall aim of this study is to evaluate the efficacy of an evidence-based MWC skills training program for improving safe MWC mobility in people with ARSACS and MD1. The primary objective is to test the hypothesis that participants who receive MWC skills training will improve their MWC skills performance by at least 20% compared to those in a control group who receive no training. The secondary objectives are to explore the influence of MWC skills training on MWC skills capacity, MWC use self-efficacy and MWC mobility. Finally, participants' expectations regarding MWC use, and experiences and perceived impact of MWC skills training on mobility and quality of life will be explored using qualitative inquiry.

Methods

Design

Sequential mixed-methods (randomized controlled trial (RCT) and qualitative interviews) will be used, with the RCT reported according to the CONSORT guidelines [27]. Participants will be randomly assigned to the intervention or control group using a 1:1 allocation ratio. A randomization process with undisclosed block size will be created by a statistician who is not otherwise involved in the project. Given the potential benefits of the intervention already reported in our previous studies [23,24], control group participants will also receive WC skills training following a 4-week control period and post-treatment data collection (i.e., they will be on a "wait-list" for the WC skills training program and receive no intervention during the study period).

Ethics Approval

The protocol for this study was approved by the Research Ethics Boards in rehabilitation and social

integration of the *Centre intégré universitaire de santé et de services sociaux de la Capitale-Nationale* (CIUSSS-CN; REB number 2025-3100). The study protocol was registered: NCT06596850. Informed consent will be obtained from all participants by a research coordinator (RC). Participants will be made aware that they can withdraw from the study at any moment without facing any consequences. All collected information will remain strictly confidential within the limits established by law. To protect privacy and confidentiality, participants will be identified by a unique code, and their data will be anonymized and securely stored on a password-protected server. Participants will receive financial compensation for data collection activities to acknowledge their time and reduce attrition (\$ 50 after each assessment session, for a total of \$150).

Participants and recruitment.

Convenience sampling will be used to recruit 20 community-dwelling people with a ARSACS or MD1 who use MWCs. Active recruitment strategies will include recruiting through outpatient services at the local rehabilitation centre in Quebec City (CIUSSS-CN). The chief of physiatry (XR) who has clinical expertise in ARSACS and MD1 will discuss the study with all individuals on his case load who have a diagnosis of ARSACS or MD1 and who use MWCs. In addition, participants from a longitudinal study on ARSACS (completed in 2022) will be contacted and informed of the study. Passive recruitment strategies will include dissemination of promotional videos on social media and websites of Muscular Dystrophy Canada and other collaborators and community organizations. Participants will be 18 years of age or older; have a diagnosis of ARSACS or MD1; use a MWC for mobility (≥ 3 times/week for ≥ 8 hours/week); and able to propel their MWC at least 10 meters. Individuals will be excluded if they anticipate a health condition or procedure that contraindicates training (e.g., surgery); or are concurrently or planning to receive other MWC training during the study.

Sample size

The sample size was powered to detect a statistically significant difference of 20% between groups using mean change scores from baseline to post-intervention in WC skills [28]. A large effect size (Cohen's $d = 1.2$) for the WST and a correlation between measures (R^2) of 0.6 was used. With $\alpha=0.05$, and 90% power for between-factors repeated measures analysis of covariance (ANCOVA) (F-tests) controlling for baseline score (account for function, WC experience), a sample size of 18 is required (calculated using G*Power [29]). Adjusting for a 10% loss to attrition, a total sample of 20 is required (10 per group).

Procedures

Following enrolment, participants will complete baseline measures (T1) and then be assigned to the experimental or control group. A RC will then contact the statistician via telephone or email to obtain group assignment within 48 hours. The participant will then be informed of group assignment, and the trainer will be given the participant's contact information to schedule the training sessions and for control group assignments, training will be scheduled after completion of blinded post-intervention data collection (T2). In effort to minimize bias, a research assistant (RA) will be blinded to group allocation and participants will be instructed not to discuss their program with the RA. The RC will also arrange for blinded follow-up (retention) data collection for all participants 3 months after post-treatment data collection (T3). Qualitative interviews will be conducted by the RC by phone or videoconferencing within 7 days of completing the intervention. Given the self-report nature of the primary outcome, that three study investigators have interdisciplinary experience with RCTs and are not located at the data collection site, and that data collection, analysis and interpretation will be discussed with a citizen partner and Muscular Dystrophy Canada, a data monitoring committee was not established. Participants in the intervention and control group will continue to receive usual care (if any) during the study.

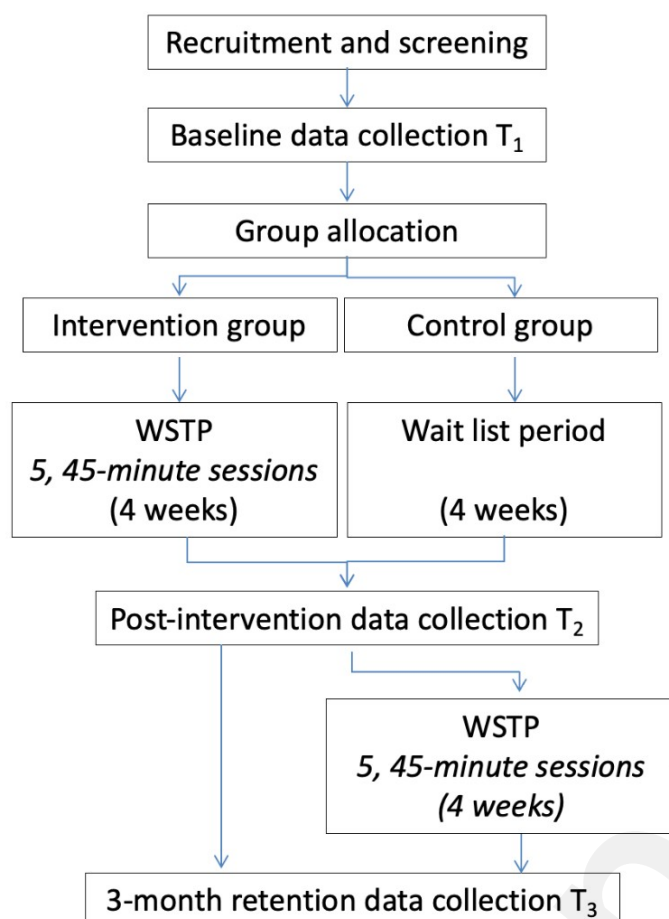


Figure 1: Trial design describing coordination, testing and experimental procedures.

Intervention

According to the evidence-based Wheelchair Skills Training Program (WSTP) [30], participants will receive 5, individual, 45-minute weekly WC skills training sessions. As previously described by Best et al., training sessions will take place in the community, in and around the participants' homes and neighborhoods (e.g., libraries, shopping centres, museums, parks) and will be customized according to participant goals [27]. The WSTP provides suggestions for individualizing the approach based on the learner's impairments (i.e., weakness, spasticity, movement disorders [like ataxia, tremor, chorea], contractures, cognition). Therefore, specific training considerations can be made regardless of diagnosis. The WSTP sessions will begin with a 5-minute review of goals/progress, followed by a 10-minute warm-up (wheeling activities, random practice of previously learned skills); 20 minutes of

attempting new skills (training on each skill will be carried to next session until the skills are learned or until the trainer and participant mutually agree that training should be abandoned; the trainer will periodically ask the participant to practice newly learned skills to incorporate variability of practice); 10 minute cool-down, during which the participant will practice skills in a self-controlled environment.

The trainer, a graduate student with training in physiotherapy (EN), will receive 2, 4-hour sessions by experts in MWC skills training (KLB, FR, RLK), which has been shown to improve knowledge and confidence for training WC skills [31]. The trainer will also complete self-guided learning modules, semi-structured practice with trained trainers, and the Wheelchair Skills Programs Certification Exam [www.wheelchairskillsprogram.ca]. Fidelity of the WSTP administration will be evaluated by a member of the study team not involved in testing or training, who will attend one training session per participant (randomly and at various times throughout the training period). The trainer will also complete a detailed training log, a checklist of standardized steps to be followed during training, and document adherence to the intervention and any adverse events. Formative assessments with the trainer will ensure fidelity.

Control group

Participants in the control group will receive usual care (if any) during a 4-week period. During this time, participants will be considered on a waitlist to receive the WSTP after the completion of the project and can continue or start any activities provided by therapists or in the community. After completion of T2 evaluations, participants in the control group will complete a target of 5, 45-minute weekly sessions of the WSTP, delivered by trained personnel as described for the intervention group. Acknowledging that usual care may include some form of MWC skills training, previous evidence suggests the amount of training received during usual care according to the WSP will be limited [25].

Data collection

The RA, an occupational therapist with 12 years of experience in clinical research, will collect quantitative data and enter them into a secure database. Sociodemographic information will include age, sex, gender, diagnosis, length of time using WC, previous experiences with using a MWC (where and for which activities), previous WC training, assistance required for using a MWC (General Scale for the Extent of Caregiver Assistance (5-point scale; no assistance (5) always autonomous, standby assistance only (4), verbal assistance only (3), 1-person physical assistance (2), 2-person physical assistance (1), equipment needed (0): available from www.wheelchairskillsprogram.ca), and propulsion method), and WC specifications (frame type, dimensions, tire and caster size).

Primary outcome

MWC skills performance (i.e., what the person does in their daily life) will be assessed using the Wheelchair Skills Test Questionnaire for MWC users (WST-Q-M v5.4.2) [32,33]. The WST-Q-M comprises 30 discrete MWC skills required to move around in a MWC indoors and in the community. Participants will rate their performance for each of the 30 MWC skills on a four-point scale. The root performance question asks, “Could you do this (MWC skill) in your own setting”. Response options include “Yes, very well (3)”, “Yes, but not well (2)”, “Yes, with help (1)”, “No (0)” and “No part (NP)” with this MWC. The total percentage WST score is calculated as: $\text{Total percentage WST Performance} = \frac{\text{sum of individual skill scores}}{([\# \text{ of possible skills} - \# \text{ of NP scores} - \# \text{ of Testing Error (TE) scores}] \times 3)} \times 100\%$. Participants will be asked whether the skill represents a training goal (yes, no) to assist with defining training program. The WST-Q can be completed in approximately 15 minutes, has strong psychometric properties in English and French [34,35], and has been used extensively in clinical trials [16]. The WST-Q-M has been validated by the study team for ARSACS [36]. A minimally clinical important difference (MCID) of 20% relative improvement in WST scores (T1 to T2) has been suggested for adults [23]. However, a recent invited commentary by

Best and Kirby suggest that the potential clinical significance of much smaller improvements should be considered [37].

Secondary outcomes

MWC skills capacity (i.e., what a person can do in a standardized environment) will be assessed using the WST Version v5.4 for MWC users [38,39]. Participants will be asked to execute 30 MWC skills in a standardized environment. Participants will be scored on a four-point scale as “Advanced pass (3)”, “Pass (2)”, “Partial pass (1)”, “Fail (0)”, “No part (NP)”, or “Testing error (TE)”. The total percentage WST score is calculated as: $\text{Total percentage WST Capacity Score} = \frac{\text{sum of individual skill scores}}{(\text{\# of possible skills} - \text{\# of NP scores} - \text{\# of TE scores}) \times 3} \times 100\%$. The WST has been validated in adult populations. A MCID of 20% relative improvement in WST scores (T1 to T2) has been suggested for adults [23].

MWC use self-efficacy (i.e., belief in ability to accomplish specific tasks while using a MWC) will be assessed using the Wheelchair Use Confidence Scale for Manual Wheelchair Users Short Form (WheelCon-M). [40]. The WheelCon-M comprises 21 statements related to confidence using a MWC in activities and environments, each rated on a scale from 0 (“not at all confident”) to 10 (“completely confident”), producing a total mean score of 0 to 10 [40,41]. Responses will indicate current level of perceived confidence in six areas (i.e., navigate the physical environment in a MWC, perform activities in a MWC, problem solve, advocate for needs, and manage social situations and emotions). The WheelCon-M was validated for ARSACS [36].

MWC mobility will be measured using actigraphy, collected with a small, non-invasive, and lightweight accelerometer (Actigraph GT3X). The Actigraph GT3X contains a multidirectional accelerometer that integrates information about direction and speed to produce an electrical current

with variable magnitude and duration, and electrical current data are stored as ‘activity counts’ [42]. Time between sampling units (epochs) will be set at 15 seconds [42].

Participants will receive two actigraphs at completion of assessments (T1, T2 and T3); one will be worn on their non-dominant arm between the elbow and shoulder, and one will be placed on the rear wheel of their MWC inside a custom-made waterproof that will be installed using tie-wraps. Participants will be asked to wear the actigraphs at all times during a 7-day period, except during sleep, bathing/showering, or swimming and to record the time the actigraph was put on and taken off using a log [43]. The tester will obtain the actigraphs and log from the participants at the end of the 7-day period (either in person or by postage paid envelopes that will be provided to the participants). Actigraphy has been validated for evaluating MWC mobility with adults [44].

Qualitative interviews

Participants will complete a 45-60-minute semi-structured individual interview in person or online with the RC after completing the WSTP intervention. The interview guide will be created by the study team using an iterative process following an appropriate framework (e.g., International Classification of Functioning, Disability and Health) [32]. Questions will explore expectations with WC use, previous experiences with using a WC, and perceptions of WC skills training. In addition, we will explore perceptions of the WSTP (dosage, fatigue, training approaches, usefulness), experiences with MWC service provision, satisfaction with MWC use and social participation. The interview guide will be pilot tested with one person who has ARSACS and one person with MD1. Interviews will be audio recorded.

Data analysis

Summary statistics (means, standard deviations, frequencies, percentages) will be used to describe the

sample. Details of adherence and intervention fidelity will be summarized. Assumptions for parametric testing of quantitative data will be verified and data will be screened for outliers [45]. Intention-to-treat analysis will be prioritized (with reporting of a per protocol analysis depending on adherence and fidelity), and missing data will be treated using multiple imputation methods for repeated methods of longitudinal data [46].

To address the primary objective, T2 WST performance scores will be compared between the experimental and control group using ANCOVA or non-parametric equivalent, with baseline scores used as the covariate. Variance (R^2), statistical significance (95% confidence interval), and effect sizes (partial η^2) will be assessed using Sums of Squares methods. Secondary objectives will be evaluated using linear mixed-effect models, which are flexible to explore within-subject mixed/random effects [47].

Post-hoc exploratory analyses will investigate the influence of age, gender, diagnoses (ataxia or dystrophy) and MWC experience on effect size using linear mixed-effect models. Comparisons between WST-Q-M within-subject change scores for the experimental group (T1-T2) and WST-Q-M within-subject change scores after training in the control group (T2-T3) will be explored using t-tests. Sensitivity analyses will be conducted on demographic variables (e.g., age, sex, gender, diagnoses) to generate hypothesis and inform future investigation.

A reflexive thematic analysis will be used to explore perceptions of the influence of MWC training on mobility, participation, and quality of life according to an appropriate framework (e.g., International Classification of Functioning, Disability and Health) [48]. Interviews will be transcribed verbatim, and researchers will familiarise with the data by re-reading and re-listening to interviews. Using NVivo software [Lumivero, 2023, Version 14] transcripts will be coded line-by-line, codes

will be grouped into similar ideas, and then themes will be developed using an inductive approach. Member checking will be performed with participants throughout data analysis, with the citizen partner actively involved in data interpretation and synthesis to enhance confirmability. Final themes and sub-themes will be verified with participants and the research team. To further enhance trustworthiness according to Ahmed (2024), credibility of data interpretation will be strengthened through triangulation with quantitative data [49]. Quantitative and qualitative data will then be integrated using joint display diagrams to merge, compare, and relate findings, to better understand how subjective perceptions align with measured improvements [50]. We will also explore why some participants may experience better outcomes or how people deal with challenges. Additionally, we will remain aware of potential personal biases regarding our perception of the effectiveness of the intervention. Finally, findings on the facilitators and barriers to training may be transferable to individuals with other neuromuscular disorders who share similar symptoms and functional characteristics. [49].

Results (Progress to date)

Research ethics was approved in July 2024. Study teams members have met to plan recruitment which will start in February 2025. A graduate student, RA, and RC have been recruited and trained. Data collected will begin in February 2025 and anticipated completion is February 2026.

Discussion

A need for improved MWC skills assessment and training in ARSACS and other neuromuscular disorders was observed and documented by our team. Given strong documented evidence among heterogenous populations of adults [23, 24], we hypothesize that an evidence-based standardized

MWC skills training program will improve MWC skill performance for people with hereditary degenerative disorders such as ARSACS and MD1. This paper describes the protocol for a randomized wait-list controlled trial to evaluate the efficacy of the WSTP on MWC performance, in adults with diagnoses of ARSCAS or MD1, and to explore the influence of training MWC skills capacity, MWC use self-efficacy, MWC mobility, and satisfaction with participation.

The United Nations declared mobility a basic human right [51]. For people with neuromuscular and hereditary degenerative disorders such as ARSACS and MD1, MWCs are commonly provided to facilitate self-directed mobility when walking becomes difficult or not possible. Just like riding a bike, using a MWC requires skill development and practice over time in indoor and outdoor environments and during real-life situations. ARSACS and MD1 are highly prevalent in the province of Quebec. This geographic distribution has positioned our team to explore longitudinal aspects of mobility and participation in these two specific disorders, which may be generalized in the future to other neuromuscular disorders (ex. Friedreich's ataxia, muscular dystrophy).

Previous studies have documented the clinical significance of learning just one MWC skill, which could mean the difference between leaving the house independently to get groceries or relying on another person to complete this instrumental activity of daily living [28]. For example, learning how to do a transient tip (i.e., popping the small front caster slightly off the ground to get over a door threshold, may make the difference of being able to leave the home or not). Once in the community, learning various approaches to opening heaving doors, manoeuvring within small spaces, and navigating irregular surfaces and curbs may make the difference in being able to go to school independently, use public transit, participate in the workforce, or socialize with friends. Given that people with ARSACS have been shown to have very low MWC skills compared to other adult MWC users (16), this study has the potential to have major implications on mobility, participation in social

and vocational activities, and quality of life for people with ARSACS and MD1.

Establishing effect size in a RCT will position the research team to work with clinical partners to implement the MWC skills training and evaluation into clinical practice. Given the proximity of researchers and clinicians involved in this study (i.e., the research centre and rehabilitation centre where outpatient services are provided are in the same building), this trial has the potential to catalyze change in MWC service provision for people with ARSACS and MD1. Implementation of MWC skills training and evaluation into clinical practice would ensure training starts early, specifically in the fundamental skills (e.g., propulsion and maneuvering) and indoor skills that are required for getting around in the home, at school and at work. Training of various approaches needed for community and advanced skills (ex. ascending curbs and steep ramps) may be customized based on disease progression, MWC configuration, and personal experiences. This in turn could reduce risks of acute (e.g., tips and falls) and overuse (e.g., repetitive strain due to poor propulsion techniques) injuries [52-54]. Booster sessions may be provided to people on an as-needed basis, such as after accidents, changes in MWCs (a service covered by the provincial health care system in Quebec every 5 years). Moreover, optimizing MWC skills may reduce caregiver burden among those who assist with mobility to accomplish activities of daily living and social participation, roles that are commonly filled by parents, spouses, and other family members. Future studies may explore facilitators and barriers to implementing MWC skills evaluation into practice with clinicians and managers.

Finally, the collaborations with clinicians, a special interest group in neuromuscular disorders (Muscular Dystrophy Canada) and a citizen partner who has lived experiences with ARSACS and MWC training research will enhance credibility of the research findings and increase likelihood of successful future implementation. New research questions may be developed based on the needs

identified by these groups.

A multifaceted knowledge translation strategy will be used to increase awareness and knowledge to change practice for MWC skills training for people with ARSACS and DM1. Targeted materials for healthcare professionals will include infographics, podcasts and video capsules that will be launched through the international PROSPAX initiative on recessive ataxias and NMD (Gagnon), including the Rare Disease International Day. Materials will be shared through clinical and community platforms including websites and social media of MDC, GRIMN, the Wheelchair Skills Program. Lunch-time presentations and MWC training workshops will be planned with clinicians. Academic dissemination will include a minimum of two peer-reviewed articles and two national/international conferences. Led by corresponding author (KB), all study investigators will have access to the final data set and will be eligible for authorship. Denominalized data will be made available to the public upon request to the corresponding author (KB).

There are limitations of this study that should be considered. Although the sample size is power for the primary outcome, we do not have power to detect changes in secondary outcomes or for subgroup analysis to explore the influence of potentially confounding factors. Considering the population under study, there may be increased risk of attrition due to fatigue or difficulty to schedule training/testing sessions within the desired timeframe. We hope to mitigate these issues by offering validated questionnaire versions of tests and by providing training in the community to overcome transportation challenges associated with coming to a research centre. Given that wheelchair configuration and positioning are outside the scope of the WSTP, we will not consider wheelchair configuration or positioning in this study. However, these factors may have an impact on MWC skills performance and capacity, and ultimately on the efficacy of the intervention.

Conclusion

The results of this randomized wait-list controlled trial will confirm whether MWC skills training may can improve self-directed mobility and related outcomes for people with ARSACS and MD1. Findings may help guide clinical practice toward MWC skills training for understanding potential influences on MWC mobility of people living with neuromuscular disorders.

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Author contributions

E Niyomwungere: Methodology, Writing - Original draft; F Routhier: Conceptualization, Methodology, Writing - Review & Editing; C Gagnon: Conceptualization, Methodology, Writing - Review & Editing; RL Kirby: Conceptualization, Methodology, Writing - Review & Editing; XRodrigue: Conceptualization, Methodology, Writing - Review & Editing; I Lessard: Conceptualization, Methodology, Writing - Review & Editing; J Lettre: Conceptualization, Methodology, Writing - Review & Editing; KLBEST: Funding acquisition, Conceptualization,

Methodology, Supervision, Projects administration, Writing - Original draft, Writing - Review & Editing

Conflicts of interest

The authors have no conflicts of interest to declare.

Abbreviations

ANCOVA: analysis of covariance; ARSACS: autosomal recessive spastic ataxia of Charlevoix-Saguenay; CIUSSS-CN: Centre intégré universitaire de santé et de services sociaux de la Capitale-Nationale; MCID: minimally clinical important difference; MD1: myotonic dystrophy type 1; MWC: manual wheelchair; NP: no part; RA: research assistant; RC: research coordinator; RCT: randomized controlled trial; TE: testing error; WC: wheelchair; WheelCon-M: Wheelchair Use Confidence Scale for Manual Wheelchair Users Short Form; WSP: Wheelchair Skills Program; WST: Wheelchair Skills Test; WSTP: Wheelchair Skills Training Program; WST-Q-M: Wheelchair Skills Test Questionnaire for MWC users

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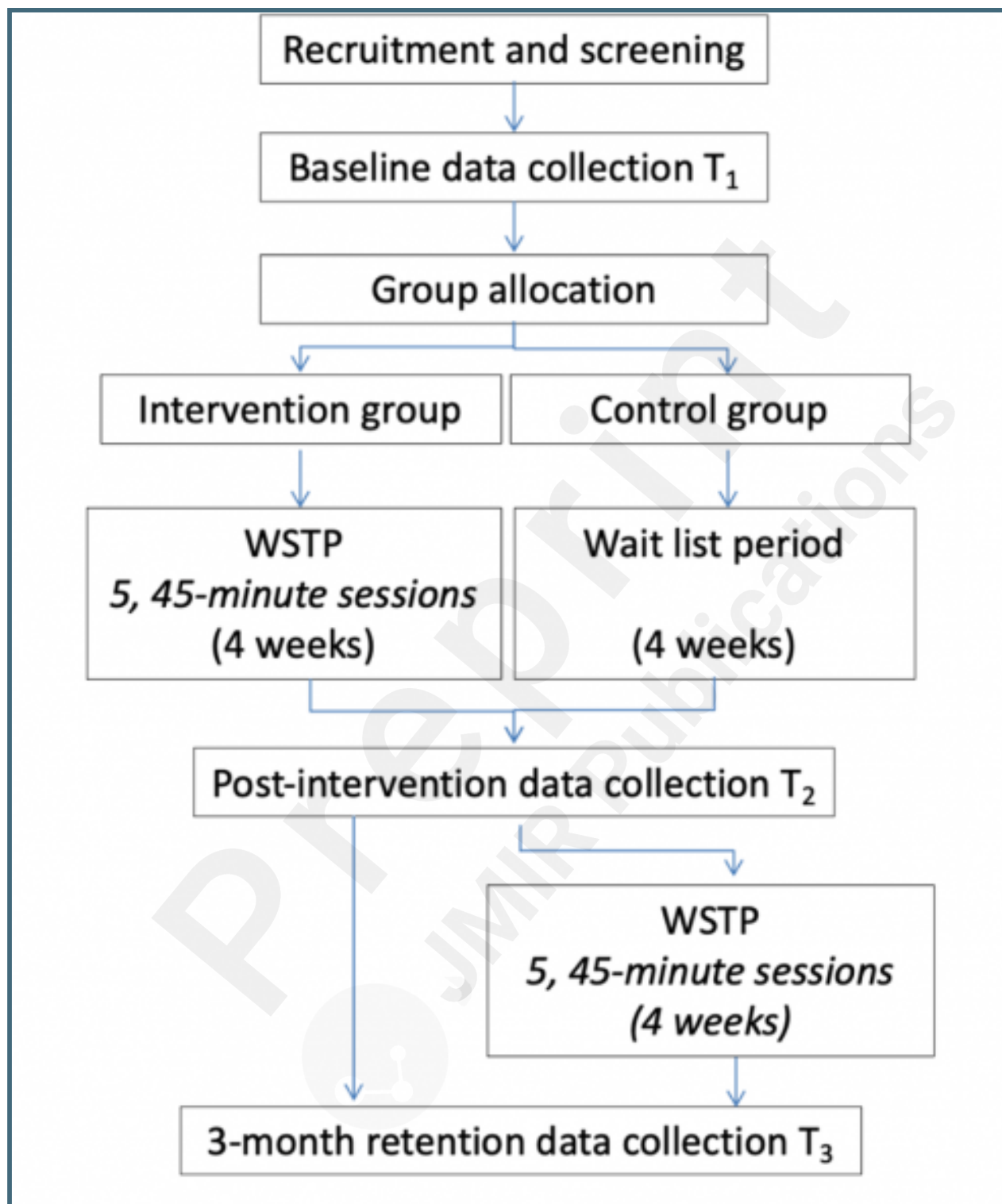
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Supplementary Files

Figures

Trial design describing coordination, testing and experimental procedures.



Multimedia Appendixes

Peer-review report.

URL: <http://asset.jmir.pub/assets/f00944c44b478774be1ed2d0bd5460c9.pdf>



CONSORT (or other) checklists

SPIRIT checklist.

URL: <http://asset.jmir.pub/assets/94ba9416236f440018455907148af844.pdf>