Taylor & Francis
Taylor & Francis Group

European Journal of Physiotherapy

ISSN: (Print) (Online) Journal homepage: https://www.tandfonline.com/loi/iejp20

The feasibility of an upper limb strength test protocol in children with Duchenne muscular dystrophy

Linda Eikeland, Eli Narum, Liv Heide Magnussen, Stian Hammer & Tiina Maarit Andersen

To cite this article: Linda Eikeland, Eli Narum, Liv Heide Magnussen, Stian Hammer & Tiina Maarit Andersen (12 Sep 2023): The feasibility of an upper limb strength test protocol in children with Duchenne muscular dystrophy, European Journal of Physiotherapy, DOI: 10.1080/21679169.2023.2256371

To link to this article: https://doi.org/10.1080/21679169.2023.2256371

© 2023 The Author(s). Published by Informa UK Limited, trading as Taylor & Francis Group.

Published online: 12 Sep 2023.

Submit your article to this journal

Article views: 321

View related articles

View Crossmark data

Full Terms & Conditions of access and use can be found at
https://www.tandfonline.com/action/journalInformation?journalCode=iejp20

EUROPEAN JOURNAL OF PHYSIOTHERAPY
https://doi.org/10.1080/21679169.2023.2256371

Taylor & Francis
Taylor & Francis Group

RESEARCH ARTICLE

OPEN ACCESS Check for updates

The feasibility of an upper limb strength test protocol in children with Duchenne muscular dystrophy

Linda Eikeland[a], Eli Narum[a], Liv Heide Magnussen[a], Stian Hammer[a,b] and Tiina Maarit Andersen[a,c]

[a]Department of Health and Function, Western Norway University of Applied Sciences, Bergen, Norway; [b]Department of Physiotherapy, Haukeland University Hospital, Bergen, Norway; [c]Department of Thoracic Medicine, Haukeland University Hospital, Bergen, Norway

ABSTRACT

Question: This study aimed to develop and assess a protocol for evaluating upper limb strength and its association with functional abilities in children and adolescents diagnosed with Duchenne muscular dystrophy (DMD).

Design: A cross-sectional study.

Participants: Eleven male individuals diagnosed with DMD (aged 11 to 18 years).

Intervention: A systematic protocol for assessing the maximum voluntary isometric contraction (MVIC) of the upper limbs using grip and pull dynamometers was developed and conducted in conjunction with the Performance of Upper Limb (PUL) scale.

Outcome measures: Feasibility was evaluated by appraising the number of participants capable of successfully executing the strength test protocol. Correlations were conducted to examine the relationship between upper limb strength and PUL scores.

Results: The grip strength assessment was feasible for all participants, while the complete strength testing protocol was feasible for nine, excluding two individuals with non-ambulatory mobility. Significant correlations were found between overall upper limb strength and total PUL scores (r_s = 0.742, $p = 0.022$), grip strength and distal PUL scores (r_s=0.733, $p = 0.010$), shoulder abduction strength and PUL shoulder scores (r_s=0.905, $p = 0.005$), and grip strength and overall PUL scores (r_s=0.794, $p = 0.004$). The middle-level correlation was not statistically significant (r_s=0.590, $p = 0.094$).

Conclusion: The established strength test protocol demonstrated feasibility among ambulatory participants. However, alternative approaches are essential for those with limited ambulation. The study identified a robust correlation between upper limb strength and functional abilities. Further extensive studies are required to validate these findings.

Trial registration: The present study is a part of a longitudinal intervention study registered at the ClinicalTrials.gov (NCT03963453).

ARTICLE HISTORY
Received 5 May 2023
Revised 25 August 2023
Accepted 3 September 2023
Published online 13 September 2023

KEYWORDS
Muscle strength; neuromuscular disease; physical function; upper extremity; dynamometer

Introduction

Duchenne muscular dystrophy (DMD) is a severe progressive neurodegenerative disease, mainly affecting boys from age 2–3 years. The loss of dystrophin degenerates the muscle fibres and leads to progressive muscle weakness [1]. Typically, muscle weakness progresses in the proximal before the distal musculature, and in the lower before the upper limbs (UL). The disease progresses fast, commonly leading to wheelchair dependency from age 12 [2]. There is no curative treatment, but glucocorticoid drug therapy has shown benefits regarding muscle strength, motor- and pulmonary function for individuals with DMD [3]. Today, survival up to the age of 30–40 years is not uncommon [2].

There is an increasing interest to evaluate the benefit of physical activity and training regimes to maintain and/or slow down the progressive loss of function, well-being and muscle strength in individuals with DMD [4]. This addresses the urgent need for standardised assessment tools to measure both strength and function in disease management and monitor disease advancements. There are several assessment tools used in children with DMD, such as the Six Minute Walk Test, North Star Ambulatory Assessment, and the Performance of Upper Limb, all measuring general function (PUL) [5]. As progressive muscle weakness is a part of the natural course of DMD, change in muscle strength can be used to monitore progression and the efficacy of therapeutic approaches. Measurements of elbow flexion- and grip strength have been recommended [5].

Individuals with DMD are easily fatigued [6] and physical testing should therefore be minimised. Given an association

CONTACT Tiina Maarit Andersen Tiina.maarit.andersen@hvl.no; Tiina.andersen@helse-bergen.no Department of Health and Function, Western Norway University of Applied Sciences, Bergen, Norway.
Linda Eikeland and Eli Narum will have a shared co-first authorship, as they have worked together and contributed equally on the manuscript.

© 2023 The Author(s). Published by Informa UK Limited, trading as Taylor & Francis Group.
This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. The terms on which this article has been published allow the posting of the Accepted Manuscript in a repository by the author(s) or with their consent.

between UL strength and physical function, muscle strength testing could in some cases replace more comprehensive functional testing. Demir et al. [7] found a moderate to strong correlation between UL muscle strength and Performance of Upper Limb (PUL) score. This however, was not the case for grip strength [7,8].

In order to gain insights into the impact of muscle strength on functional performance in this specific patient population, and to explore whether these measures could serve as an acceptable substitute for more comprehensive functional assessments the following aims were formulated; to evaluate the feasibility of a newly developed standardised testing protocol designed to assess UL muscle strength within the paediatric and adolescent DMD population, and secondly to explore the correlation between UL strength and UL function.

Material and methods

Study design

This cross-sectional study assessed a regional population of individuals with DMD in March/April 2021. All participants provided written informed consents. The study was part of a longitudinal intervention study registered under ClinicalTrials.gov Identifier: NCT03963453 and approved by The Western-Norway Regional Committee of Medical Research Ethics (2019/00260).

Subjects

All ($n = 12$) individuals with DMD followed up at the paediatric rehabilitation clinic at a Norwegian hospital, were invited to participate in the study. Inclusion criteria were: DMD diagnosis, 6–18 years, and ability to give informed consent, perform physical exercises, and to understand instructions. Exclusion criteria were being too weak to perform the standardised test protocol correctly, language barriers, cognitive dysfunction, or inability to understand instructions.

Materials and procedure

Demographic and clinical variables

Descriptive characteristics regarding the participants age, weight, height, body mass index (BMI), DMD stage, Brooke scale, Vignos scale and use of glucocorticoids, were gathered from the medical journals.

Maximum voluntary isometric contraction (MVIC)

The strength test protocol (Tables 1 and 2) was developed in collaboration with a multidisciplinary team and with feedback from international DMD scientists, and user representatives in the present study. The protocol was pilot tested on healthy children and children with DMD to make appropriate adjustments regarding physical impairments and contractures, and to increase reproducibility.

A grip dynamometer (KFORCE grip by KINVENT, Montpellier, France) was used to measure grip strength. A force transducer (pull dynamometer) (KFORCE Link by KINVENT, Montpellier, France) was used for investigating maximum voluntary isometric contraction (MVIC) of shoulder abduction, elbow flexion and -extension. The equipment was connected to a mobile application (KFORCE by KINVENT, Montpellier, France) displaying a curve of strength development (Kg/s) and peak torque (Kg). The participants were tested on an adjustable treatment table in supine position. Three attempts were performed on both right and left side, with three seconds MVIC for each test, and 20 s rest between the attempts. If the participant was unable to perform a test in standardised position, a deviation was noted. If the participant deviated from the protocol, the attempt was regarded as invalid, and a new attempt was offered. Challenges observed during the testing were registered. The test protocol took approximately 20–30 min to complete. The strength test protocol is not evaluated for reliability or validity.

The feasibility of the strength test protocol was defined as the ability to complete the tests according to the protocol. Minor deviations in positioning due to contractures, were still regarded as valid. Attempts were considered invalid and excluded from analysis, if the participant had a peak torque less than the weight of the equipment (1.2 kg). Test trials not following the standardised test protocol were also excluded.

For the participants safety, pain and perceived exertion were registered.

The Numeric Rating Scale (NRS) ranging from 0to 10 (where 0 is no discomfort and 10 worst possible discomfort) was used to report perception of pain during testing. NRS has been found acceptable for self-reported acute pain in children [10].

A Norwegian version of the 'Children's OMNI scale of perceived exertion' was used to evaluate the participants perceived exertion after completion of the strength protocol. The scale is validated for use in children performing resistance exercise [9].

Performance of upper limb (PUL)

The study employed version 2.0 of the Performance of Upper Limb (PUL) assessment tool to quantify UL function [11]. PUL 2.0 represents a streamlined iteration of its predecessor, PUL 1.2, which possesses established credibility in the evaluation of UL function across both ambulant and non-ambulant individuals with DMD [12]. Notably, both versions have demonstrated a high degree of sensitivity in detecting changes across all tiers of assessment [13]. This evaluative protocol encompasses three distinct domains: 1) Shoulder, 2) Middle, and 3) Wrist and fingers (distal). Scoring was conducted exclusively using the dominant or preferred side of the individual. Shoulder-level tasks encompassed lifting the arm with and without weights, whereas middle-level tasks encompassed the manipulation of weights ranging from 0,1 to 1 kg, bringing the hand to the mouth, stacking cans, and opening containers. Distal-level tasks encompassed activities such as tearing paper, exerting force on a light switch, employing a pencil, lifting a 10 g weight, collecting coins, and executing arm supination. The protocol was meticulously adhered to, yielding a total possible score of 42. Moreover, sub-scores

Table 1. Equipment for the strength test protocol for children and adolescents with DMD.

Materials	Quantity	Description
Treatment table	1	For positioning the participants and adjusting height to achieve a horizontal force output.
Wall $_{bars}$	1	For fastening the end of the rope and/or the adjustable rigid strap.
Grip dynamometer (KFORCE Grip by KINVENT, Montpellier, France)	1	Measures hand grip strength.
Pull dynamometer (KFORCE Link by KINVENT, Montpellier, France)	1	Measures isometric strength.
Smartphone with the KFORCE application (by KINVENT, Montpellier, France)	1	The dynamometers connect to a mobile application by Bluetooth, containing the tests and registers test results.
Adjustable rigid strap	1	Attaches to the wall bars and the pull dynamometer to adjust the strap to the appropriate length.
Loop fastening accessory	1	Attaches the pull dynamometer to the participants.
Carabiner hook	2	Fastens the adjustable rigid strap and the loop fastening accessory to the pull dynamometer.
Rope	1	Elongates the adjustable rigid strap.
Positioning bolster	2	Placed under the knees and ankles for comfort.
Traction/fixation belt	2	To fixate the participants to the treatment table to prevent movement in the longitudinal and lateral direction. One belt placed between the legs, the other horizontally above their waist.
Towel	1	Placed underneath the fixation belts for comfort.
Pillow	1	Supporting the participant's head.

were determined, allocating a maximum of 12 to the shoulder domain, 17 to the middle domain, and 13 to the distal domain [13]. Qualified physiotherapists, endowed with substantial experience in PUL administration, conducted video recorded PUL assessments. Instances of score disagreement were addressed through collaborative consultation with another physiotherapist. The testing duration approximated between 15 to 30 min.

Statistical analysis

Statistical analyses were conducted using SPSS v27.0 (IBM, Armonk, USA). Descriptive analyses were used to describe the study sample. Data are presented as means with standard deviations (SD), or frequencies (*n*) and percentages (%), as appropriate. Invalid results and missing data were excluded from analyses with a listwise deletion for the specific test. Association analyses were performed for total scores in strength and PUL, and for each subtest in PUL and muscle groups at the corresponding extremity level. The distributions of data were visually inspected through histograms and Q-Q plots for normality. Spearman's rho correlation coefficient (r_s) was used to examine the association between UL strength and PUL. The strength of the correlations was determined as used by Bulut et al. [8] and Demir et al. [7] accordingly: Very weak $r = 0.00–0.30$; Weak $r = 0.30–0.50$; Moderate $r = 0.50–0.70$; Strong $r = 0.70–0.90$; Very strong 0.9–1.00. *P*-values were two-sided and significance level was set below 0.05.

Results

Participants

Initially, the study included 12 participants. One of the participants was excluded as his physical strength was poorer than required according to the strength test protocol, leaving 11 participants for further analyses. The participants were boys from 11 to 18 years. The majority were in DMD stage 1 or 4, and five participants were non-ambulatory (wheelchair bound). All but one had received glucocorticoid treatment for more than six months (Table 3).

Feasibility

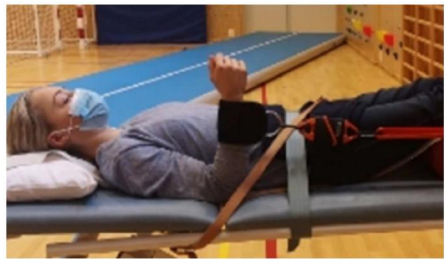
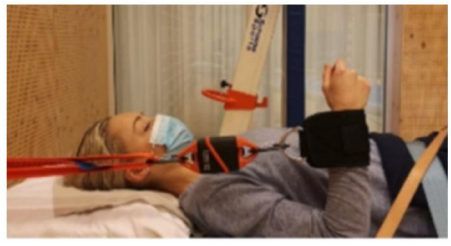
The strength test protocol was considered feasible for nine of the 11 participants. Both participants that were unable to complete the protocol were non-ambulant. One was too weak to perform MVIC elbow flexion, and the other too weak to perform MVIC elbow extension. All participants were able to perform the grip strength test regardless of the disease stage.

Challenges related to the strength testing were observed (Table 4). The elbow tests were found to be impossible to complete for some of the weaker individuals as the weight of the equipment was too heavy. The standardised test position for shoulder abduction was not ideal for participants who were overweight, as the force transducer would cause friction against their chest, despite adjusting to increased shoulder flexion (from the standard 45°to 60°). Most participants performed the test with internal rotation in the shoulder, due to contractures. There were no identifiable difficulties with the equipment regarding technological functioning, including the pairing with- and operating through the mobile application.

Association between UL strength and PUL

There was a moderate to strong-, positive linear association between strength tests and PUL for the different subtests (Figure 1). There were no clear outliers. Each scatter plot reveals a high plateau where PUL reaches maximum score values as MVIC values continue to increase. There were

Table 2. Descriptions and instructions for the strength test protocol for children and adolescents with DMD.

Testers	
Two testers were responsible for conducting the protocol and to encourage the test subject. The participants' dominant side were registered prior to testing	
Pre-instructions You will execute a sample of strength tests and be tested respectively on the right and left side. There will be three attempts on each side. A fourth attempt will be executed if your third attempt is your best. Try to perform the test with maximum strength for the duration of three seconds. There will be a 20 s rest between attempts. Start and stop cues will be given by the testers. Report any perception of pain or discomfort during the tests, using a scale from 0 to 10, where 0 is no discomfort and 10 worst possible discomfort.	
Grip strength Lying supine. The upper arm resting on the treatment table. Elbow 90° flexion. Forearm and wrist in neutral position. *Instruction* Squeeze as tight as you can. 3-2-1-start!	
Elbow flexion Lying supine. The upper arm resting on the treatment table. Elbow 90° flexion. Forearm and wrist in neutral position. Loop fastening accessory just proximal to the wrist. *Instruction* Bend your elbow as hard as you can. 3-2-1-start!	
Elbow extension Lying supine. Elbow 90° flexion. Upper arm resting on the treatment table. Forearm and wrist in neutral position. Loop fastening accessory just proximal to the wrist. *Instruction* Extend your elbow as hard as you can. 3-2-1-start!	
Shoulder abduction Lying supine. 45° shoulder flexion and 10° shoulder abduction. Loop fastening accessory just proximal to the elbow. *Instruction* Pull your arm sideways as hard as you can. 3-2-1-start!	
Registration of perceived exertion After the completion of the test protocol, a Norwegian version of the 'Children's OMNI scale of perceived exertion' was used to evaluate the participants perceived exertion [9]. They were explained that 0 is perceived as not tired at all and 10 is very, very tired, before being asked to report their level of exhaustion.	

The four pictures in the table illustrate the test positions for each subtest as described in the table text.

moderate to high statistically significant correlations between total UL strength and total PUL (r_s=0.742, p = 0.022), grip strength and PUL distal (r_s=0.733, p = 0.010), shoulder abduction strength and PUL shoulder (r_s=0.905, p = 0.005), and grip strength and total PUL (r_s=0.794, p = 0.004). The correlation at middle level was not statistically significant (r_s=0.590, p = 0.094).

Discussion

The study demonstrates the feasibility of grip strength testing in individuals with DMD, alongside feasible pull dynamometer strength testing primarily among ambulatory boys. Strong, significant associations were observed between various UL strength tests and UL function.

Development of the strength protocol

Various measures were taken to increase the measurement quality and minimise error, including collaboration with a multidisciplinary team, international collaborators and user representatives. Since high intensity- and eccentric strength training are not recommended for individuals with DMD due to the potential risk of muscle damage [2], maximum

Table 3. Participant characteristics ($n = 11$).

	Mean (SD)
Age (years)	14.5 (2.63)
Weight (kg)	52 (32.91)
Height (cm)	134 (14.17)
BMI	24.4 (9.41)
DMD stage (1–4)	2.4 (1.43)
Brooke scale	1.9 (1.14)
Vignos scale	5.8 (3.13)

BMI = Body Mass Index; DMD stage: 1= Early ambulatory; 2= Late ambulatory; 3= Early non-ambulatory; 4= Late non-ambulatory. Vignos scale for lower extremities (1–10); Brook scale for upper extremities (1–6) [14].

Table 4. Observations from strength testing.

Subtest		Observations
Grip strength	+	All participants were able to execute the test according to the test battery.
Elbow flexion and -extension	+	All participants were able to attain the standardised positions
	–	Unattainable for two of the participants due to their limitation in strength.
Shoulder abduction	–	Not ideal for overweight participants
	–	Most participants were unable to attain the standardised position of 0° shoulder rotation.

voluntary isometric contraction (MVIC) strength testing was selected. Muscle groups were selected based on recommendations for the DMD population [5] and previous studies [7,15–17]. Grip strength was included as it is feasible in DMD [5], is indicated to reflect overall muscle strength, and to have clinical and prognostic value [18].

Both the handheld dynamometer (HHD) and the pull dynamometer were under consideration for the strength test protocol. While the HHD has been utilised for assessing UL strength in children with DMD in previous studies [16,19], the decision to employ the pull dynamometer was driven by concerns related to the HHD's substantial tester influence [20] and its inclination to overestimate muscle weakness in individuals with neuromuscular disorders [21]. The selection of the pull dynamometer was reinforced by the findings of preliminary testing, which indicated reduced outcome variability compared to the HHD. The determination of optimal test positions was informed by the insights gained from preliminary testing, taking into account considerations of physical function and impairments. Comprehensive descriptions of these positions were provided to mitigate potential sources of measurement error.

Feasibility

Grip strength proved feasible for all participants, aligning with its ease, fast and non-invasive nature, rendering it suitable regardless of DMD stage. These findings concur with Birnkrant et al.'s [5] recommendation that grip strength assessment is feasible in DMD. The pull dynamometer exhibited feasibility in assessing UL strength among ambulant DMD participants, except in two non-ambulant individuals. Similar outcomes were seen in Elsheikh et al. [22], who studied ambulant adults having spinal muscular atrophy, where pull dynamometer testing was feasible. Conversely, our PUL results unveiled a ceiling effect in stronger ambulant participants, supporting earlier indications that PUL is most relevant for non-ambulant DMD individuals [5].

Insights gained during testing unveiled challenges within the strength test protocol. Elbow tests were unfeasible for weaker non-ambulant participants due to the equipment weight and alternative methods are needed. The shoulder abduction strength assessment was suboptimal for overweight participants due to friction from the force transducer, accentuating its unsuitability for the overweight DMD population, compounded by weight gain risk [1]. An internal rotation pattern in the shoulder was common, reflecting the impact of contractures [1], necessitating their consideration in future tests. Most of the participants executed this test with internal rotation in the shoulder due to contractures limiting their ability to perform the test with 0° rotation. Contractures are a normal part of the disease progression [1] and must be regarded when developing future tests.

Association between strength and function

Our study's UL strength-function association mirrors the results of Demir et al. [7], showing a moderate- to strong correlation between UL strength and total PUL in non-ambulant individuals. However, the robust grip strength total PUL association uncovered in the present study has not been noted in non-ambulant DMD individuals before [7,8]. A potential explanation is that the proximal domain subtests within PUL were significantly compromised for most participants [8]. Subtests in other domains, unlinked to grip function, were less demanding and could therefore be performed without excessive effort. In the present study, both ambulant and non-ambulant individuals participated, and the majority was still able to perform several PUL shoulder level subtests. This may indicate that the correlation of grip strength with overall UL physical is comparatively less robust in DMD than in other populations. Notably, the grip strength-function correlation might be weaker in non-ambulant individuals, due to earlier proximal musculature weakening relative to distal muscles, and progressive contractures' influence [2]. This substantiates the suitability of grip strength solely for the ambulant subgroup in function and progression screening. However, the appropriateness of UL strength measures as UL function proxies remains uncertain.

Limitations

The UL strength test protocol lacks evaluation for reproducibility, raising uncertainty about the validity and reliability of findings. The present study was conducted at the beginning of a longitudinal one-year training intervention implying that the participants went through an extensive amount of testing procedures. Therefore, we considered that adding repeated reliability testing could negatively affect the

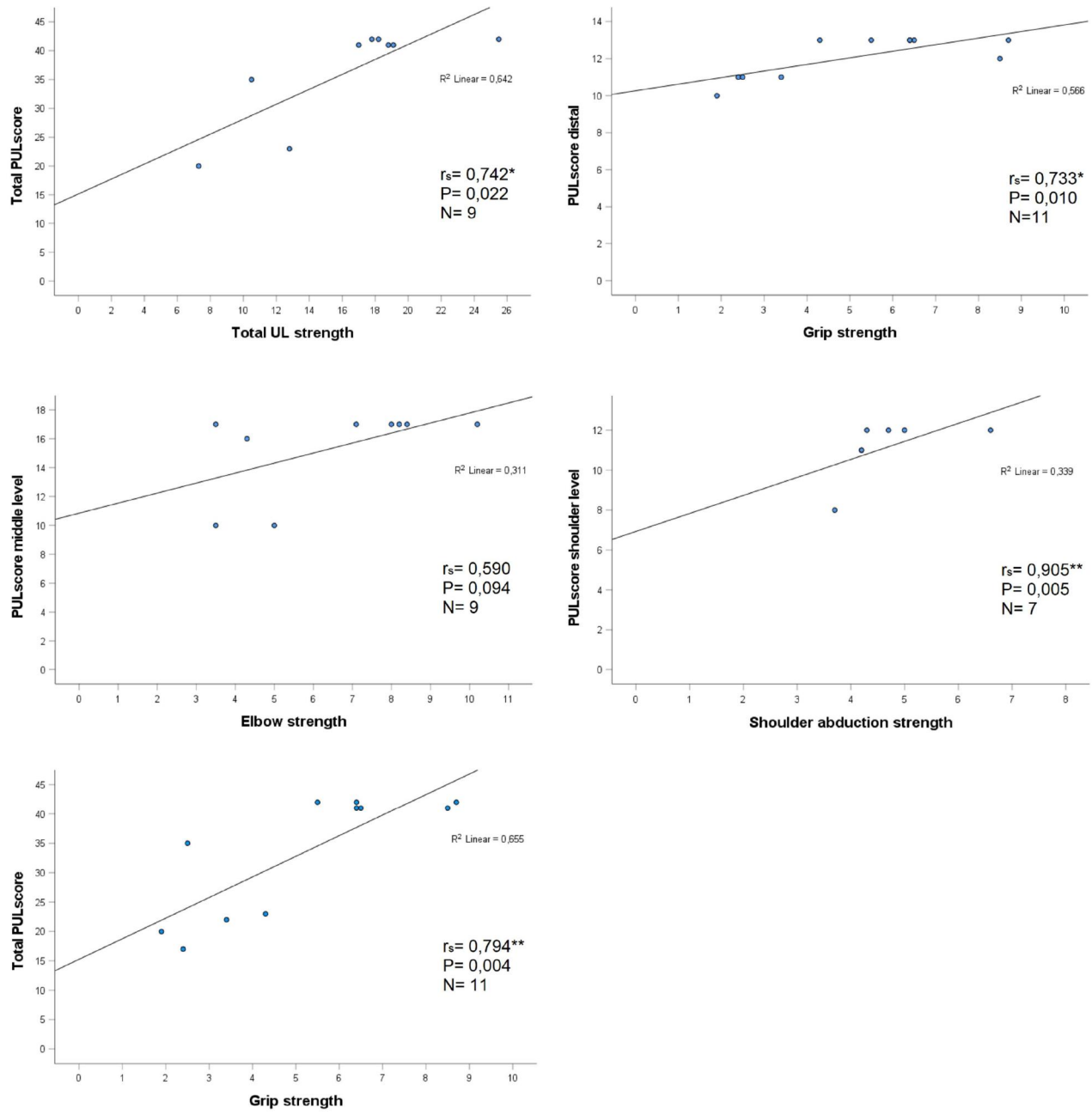


Figure 1. Scatter plots of associations between upper limb (UL) strength and performance of upper limb (PUL). *$p < 0.05$; **$p < 0.01$; r_s=Spearman's rho.

participants motivation and capacity. Moreover, in DMD, there is a risk of reproducibility bias resulting from exhaustion, or even fatigue, further raising an ethical concern due to potentially harming the muscles [5].

Reliability assessments necessitate a separate study. However, Elsheikh et al. [22] found excellent test-retest reliability using a pull dynamometer in ambulant adults with spinal muscular atrophy, suggesting reliability for other muscle weakness disorders. Further, the protocol bears time consumption and equipment adjustments, affecting motivation, especially in paediatric testing. Pull dynamometry limitations entail equipment adjustments and access to a fixed frame.

Furthermore, the researchers were neither independent nor blinded. They participated in the development of the strength test protocol and conducted the testing procedure. Yet, the testers strived to be neutral and objective to minimise potential impact on the results.

The small convenience sample is another limitation of this study. DMD is a rare disease with totally ~100 individuals in Norway [23], making it challenging to recruit a larger sample more representative to the heterogeneous population of DMD individuals [24]. However, all the individuals who met the inclusion criteria participated with no dropouts. Small sample size led to near-normal data distribution, necessitating to use low-powered Spearman's rho correlation coefficient [25]. Further research is needed, and ideally, such studies should include large groups of participants with variation in disease severity. Due to the rarity and nature of DMD, this will require international multicentre studies to include sufficient numbers.

Implications for physiotherapy practice

From a physiotherapeutic perspective, the main goal in treatment of individuals with DMD is to improve or maintain function [5]. Due to the nature of the disease, measures for minimal muscles changes are required. Muscle strength testing might replace comprehensive functional testing, crucial for fatigue-prone individuals. The present study provides a feasible UL muscle strength test protocol, notably in ambulatory individuals, necessitating alternative positions for weaker non-ambulant cases.

This study furthers understanding of UL strength- and function interplay in DMD. Differing functioning/activity levels in ambulant and non-ambulant DMD necessitate distinct measurement tools. The strength test protocol captures more score variations than PUL in better-functioning individuals, aiding physiotherapists in targeted treatment decisions based on disease stage.

Conclusion

Grip strength was feasible for all participants and the pull dynamometer was feasible in all ambulant participants with DMD. Due to the small study sample size, in addition to the strength test protocol not being evaluated for reliability, the results must be interpreted with caution and cannot be generalised to all individuals with DMD. The present study has investigated appropriate methods for assessing muscle strength using a pull dynamometer in individuals with DMD, and our findings may provide insight for developing future studies.

Acknowledgements

We extend many thanks to Bård Bogen, Guri Brekke and Donovan Lott for their valuable advices when planning the strength testing protocol. We are also very grateful to Roy Miodini Nilsen for his help with the statistics.

Ethics approval statement

The study was approved by The Western-Norway Regional Committee of Medical Research Ethics (2019/00260).

Patient consent statement

Informed consent was obtained from all subjects involved in the study.

Study registration

The present study is a part of a longitudinal intervention study registered at the ClinicalTrials.gov (NCT03963453).

Author contributions

LE, EN are contirbuted for literature search. LE, EN are contirbuted for data collection. TMA, SH, LHM, LE, EN are contirbuted for study design. LE, EN, LHM, TMA are contirbuted for analysis of data. LE, EN, LHM, TMA are helped for manuscript preparation. TMA, LHM, EN, LE, SH are review of manuscript.

Disclosure statement

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Funding

This project has been made possible by the funding received from Dam Foundation, grant number 10.13039/100009471.

ORCID

Liv Heide Magnussen http://orcid.org/0000-0001-8119-7922
Stian Hammer http://orcid.org/0000-0003-2254-7704
Tiina Maarit Andersen http://orcid.org/0000-0002-0641-3653

Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

References

[1] Munot P. 2021. Muscular dystrophies. In BMJ best practice. Available from https://bestpractice.bmj.com/topics/en-gb/969/pdf/969/Muscular%20dystrophies.pdf
[2] Darras BT. 2020a. Duchenne and becker muscular dystrophy: clinical features and diagnosis. In *UpToDate*. Available from: https://www.uptodate.com/contents/duchenne-and-becker-muscular-dystrophy-clinical-features-and-diagnosis?source=history_widget.
[3] Darras BT. 2020b. Duchenne and becker muscular dystrophy: management and prognosis. In *UpToDate*. Available from: https://www.uptodate.com/contents/duchenne-and-becker-muscular-dystrophy-management-and-prognosis?search=duchenne%20muscular%20dystrophy&topicRef=116654&source=related_link.
[4] Hammer S, Toussaint M, Vollsæter M, et al. Exercise training in duchenne muscular dystrophy: a systematic review and meta-analysis. J Rehabil Med. 2022;54:jrm00250. doi: 10.2340/jrm.v53.985.
[5] Birnkrant DJ, Bushby K, Bann CM, et al. Diagnosis and management of duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management. Lancet Neurol. 2018;17(3):251–267. doi: 10.1016/S1474-4422(18)30024-3.
[6] Angelini C, Tasca E. Fatigue in muscular dystrophies. Neuromuscul Disord. 2012;22(Suppl 3–3):S214–S220. doi: 10.1016/j.nmd.2012.10.010.
[7] Demir G, Bulut N, Yılmaz Ö, et al. Manual ability and upper limb performance in nonambulatory stage of duchenne muscular dystrophy. Arch Pediatr. 2020;27(6):304–309. doi: 10.1016/j.arcped.2020.06.007.
[8] Bulut N, Gürbüz I, Yilmaz Ö, et al. The association of hand grip strength with functional measures in non-ambulatory children with duchenne muscular dystrophy. Arq Neuropsiquiatr. 2019;77(11):792–796. doi: 10.1590/0004-282X20190161.
[9] Robertson RJ, Goss FL, Andreacci JL, et al. Validation of the children's OMNI-resistance exercise scale of perceived exertion. Med Sci Sports Exerc. 2005;37(5):819–826. doi: 10.1249/01.MSS.0000162619.33236.F1.
[10] Birnie KA, Hundert AS, Lalloo C, et al. Recommendations for selection of self-report pain intensity measures in children and adolescents: a systematic review and quality assessment of measurement properties. Pain. 2019;160(1):5–18. doi: 10.1097/j.pain.0000000000001377.
[11] Mayhew A, Mazzone ES, Eagle M, et al. Development of the performance of the upper limb module for duchenne muscular

dystrophy. Dev Med Child Neurol. 2013;55(11):1038–1045. doi: 10.1111/dmcn.12213.

[12] Pane M, Mazzone ES, Fanelli L, et al. Reliability of the performance of upper limb assessment in Duchenne muscular dystrophy. Neuromuscul Disord. 2014;24(3):201–206. doi: 10.1016/j.nmd.2013.11.014.

[13] Mayhew A, Coratti G, Mazzone ES, et al. Performance of upper limb module for duchenne muscular dystrophy. Dev Med Child Neurol. 2020;62(5):633–639. doi: 10.1111/dmcn.14361.

[14] Lue Y-J, Lin R-F, Chen S-S, et al. Measurement of the functional status of patients with different types of muscular dystrophy. Kaohsiung J Med Sci. 2009;25(6):325–333. doi: 10.1016/S1607-551X(09)70523-6.

[15] Alemdaroğlu I, Karaduman A, Yilmaz ÖT, et al. Different types of upper extremity exercise training in duchenne muscular dystrophy: effects on functional performance, strength, endurance, and ambulation. Muscle Nerve. 2015;51(5):697–705. doi: 10.1002/mus.24451.

[16] Heutinck L, Jansen M, van den Elzen Y, et al. Virtual reality computer gaming with dynamic arm support in boys with Duchenne Muscular Dystrophy. J Neuromuscul Dis. 2018;5(3):359–372. doi: 10.3233/JND-180307.

[17] Killian M, Buchowski MS, Donnelly T, et al. Beyond ambulation: measuring physical activity in youth with Duchenne Muscular Dystrophy. Neuromuscul Disord. 2020;30(4):277–282. doi: 10.1016/j.nmd.2020.02.007.

[18] Bohannon RW. Muscle strength: clinical and prognostic value of hand-grip dynamometry. Curr Opin Clin Nutr Metab Care. 2015;18(5):465–470. doi: 10.1097/MCO.0000000000000202.

[19] Kenis-Coskun O, Imamoglu S, Karamancioglu B, et al. Comparison of telerehabilitation versus home-based video exercise in patients with Duchenne muscular dystrophy: a single-blind randomized study. Acta Neurol Belg. 2022;122(5):1269–1280. doi: 10.1007/s13760-022-01975-4.

[20] Neumann DA. 2017. Kinesiology of the musculoskeletal system: foundations for rehabilitation. 3rd ed. St. Louis, MO: Elsevier.

[21] Brinkmann JR, Andres P, Mendoza M, et al. Guidelines for the use and performance of quantitative outcome measures in ALS clinical trials. J Neurol Sci. 1997;147(1):97–111. doi: 10.1016/S0022-510X(96)05220-3.

[22] Elsheikh B, King W, Peng J, et al. Outcome measures in a cohort of ambulatory adults with spinal muscular atrophy. Muscle Nerve. 2020;61(2):187–191. doi: 10.1002/mus.26756.

[23] Helsenorge. Duchennes muskeldystrofi (DMD). December 15; 2021. Available from https://www.helsenorge.no/sykdom/sjeldne-diagnoser/nevromuskulare-sykdommer/duchenne-muskeldystrofi/.

[24] Aalen OO, Frigessi A. 2006. Statistiske metoder i medisin og helsefag. Oslo: Gyldendal akademisk. Available from: https://www.nb.no/search?q=oaiid:"oai:nb.bibsys.no:990600710954702202"&mediatype=bøker

[25] Polit DF, Beck CT. Nursing research: generating and assessing evidence for nursing practice. 11th ed.; International edition. Philadelphia: Lippincott Williams & Wilkins; 2020.