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ARTICLE

The association of hand grip strength with functional measures in non-ambulatory children with Duchenne muscular dystrophy

Associação da força de preensão manual com medidas funcionais em crianças com distrofia muscular de Duchenne, que não deambulam

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ABSTRACT

Duchenne muscular dystrophy (DMD) is a disease characterized by progressive loss of muscle fiber, gradually from proximal to distal. Although a few studies have investigated hand grip strength in non-ambulatory DMD patients, a lack of literature was found determining its relationship with functional capacity. **Objective:** The aim of this study was to determine the associations between hand grip strength and functional measures in non-ambulatory children with DMD. **Methods:** Hand grip strength was evaluated using a dynamometer in children with DMD. The children with DMD were evaluated with the Turkish version of the Egen Klassifikation Scale Version 2 (EK2) for global functional capacity, the Performance of Upper Limb (PUL) for upper limb functional performance and the ABILHAND-Kids for hand ability. **Results:** The mean age of 38 DMD children was 12.02 ± 1.99 years. Dominant hand grip strength of the children with DMD was higher than the non-dominant hand ($p < 0.05$). The EK2 was 13.02 ± 5.50, PUL was 49.86 ± 14.34 and ABILHAND-Kids was 26.81 ± 7.59. Hand grip strength was found to be correlated with the EK2 ($p < 0.05$). **Conclusions:** It is known that measuring functional ability and strength in very weak children with DMD has been difficult and complex for therapists/clinicians in the clinical environment. Although there is a moderate correlation, hand grip strength may be used in clinical practice as a practical assessment tool to have an immediate insight into the global functional capacity in non-ambulatory DMD children.

Keywords: Neuromuscular disease; muscular dystrophy; Duchenne muscular dystrophy.

RESUMO

A distrofia muscular de Duchenne (DMD) é uma doença caracterizada por perda progressiva da fibra muscular, gradualmente de proximal a distal. Embora poucos estudos tenham investigado a força de preensão manual em pacientes com DMD não ambulatoriais, foi observada uma falta de literatura para determinar suas relações com a capacidade funcional. **Objetivo:** O objetivo deste estudo foi determinar as associações entre força de preensão manual e medidas funcionais em crianças não ambulatoriais com DMD. **Métodos:** A força de preensão manual foi avaliada com dinamômetro em crianças com DMD. As crianças com DMD foram avaliadas com a versão turca da Egen Klassifikation Scale Versão 2 (EK2) para capacidade funcional global, desempenho do membro superior (PUL) para desempenho funcional do membro superior e ABILHAND-Kids para a habilidade manual. **Resultados:** A idade média de trinta e oito crianças com DMD foi de 12,02 ± 1,99. A força de preensão manual dominante das crianças com DMD foi maior que a da mão não dominante ($p < 0,05$). A EK2 foi calculada em 13,02 ± 5,50, PUL em 49,86 ± 14,34 e ABILHAND-Kids em 26,81 ± 7,59. A força de preensão manual foi correlacionada com a EK2 ($p < 0,05$). **Conclusões:** Sabe-se que medir a capacidade funcional e força em crianças muito fracas com DMD tem sido difícil e complexo para terapeutas / clínicos em ambiente clínico. Embora exista uma correlação moderada, a força de preensão manual pode ser usada na prática clínica como uma ferramenta de avaliação prática para obter imediatamente uma percepção da capacidade funcional global em crianças com DMD não ambulatoriais.

Palavras-chave: Doenças neuro-musculares; distrofias musculares; distrofia muscular de Duchenne.

Duchenne muscular dystrophy (DMD) is a neuromuscular disease characterized by the gradually loss of strength starting from the proximal muscles[1]. It is diagnosed around the age of four years when symptoms such as fatigue, frequent

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falls, Gowers' sign and tip-toe walking are seen. Due to the progressive nature of the disease, children lose their ability to walk at about 9–12 years of age, and become wheelchair dependent in time. Children usually die in the second decade because of respiratory and cardiac complications[2,3].

Since the disease has a fatal natural progress, many clinical trials have been carried out to increase lifespan in recent years, in addition to the developments in therapeutic approaches, such as medical and physical therapy to slow down the symptoms. Physical outcome measures are the most preferred methods for indicating the efficacy of therapy options. Although there are many outcome measures, such as timed performance tests including the Six-Minute Walk Test, North Star Ambulatory Assessment, and the Performance of Upper Limb (PUL) assessment for DMD, the vast majority of assessment tools can only be performed by ambulatory children[4]. A limited number of measures have been suggested in the literature to determine the functional status of non-ambulatory children with DMD such as the Egen Klassifikation Scale Version 2 (EK2) and the Motor Function Measure for general functional ability, and the PUL for upper extremity performance. In clinical settings, therapists sometimes need more practical assessment methods to better manage the time with their pediatric patient.

Assessment of hand grip strength is often used for evaluating distal muscle strength, due to its ease of application in many orthopedic and neurologic diseases[5,6]. This measure, which is also used in pediatric neuromuscular patients, has also been reported to show distal muscle weakness as well as proximal weakness in the earliest phases of the disease[7,8]. The literature presents a limited number of studies that suggest evaluating hand grip strength to determine upper extremity functional status and to show the relationship with the ability of upper extremities in early rheumatoid and home care patients[9,10]. Although it has been shown that hand weakness was related to global muscular strength of the hand and physical level in children with DMD[11], to our knowledge, no other studies have been found in the literature that investigate whether hand grip strength is correlated with upper limb performance and general functional capacity, and whether it could be used by therapists for time efficiency in clinical practice to indicate the general functional capacity of a non-ambulatory child with DMD.

The aim of this study was to investigate the association between an easily applicable method—hand grip strength—and global and/or regional functional assessment tools for non-ambulatory children with DMD.

METHODS

Participants

Ethical approval was obtained from the Hacettepe University, Non-invasive Clinical Research Ethical Committee. Written consent was received from both the children and their families included in the study.

The inclusion criteria for the study were children with DMD who were: a) diagnosed with genetically-confirmed DMD with deletion in the dystrophin gene location; b) unable to walk; c) age 5–18 years old; and d) on corticosteroid treatment for more than six months. Children with cognitive and behavioral problems that might have impaired compliance with the assessments, who had severe contractures, and who had previous injury or surgery associated with the upper extremity, were excluded from the study.

The physical characteristics of the children with DMD such as age (years), weight (kg), height (cm), body mass index (kg/m^2), and dominant hand were recorded. The following assessments were performed after recording the information obtained from the parents of children on the duration of corticosteroid treatment and the duration of the non-ambulatory phase.

Assessment of hand grip strength

The hand grip strength of children included in the study was evaluated using the Jamar Hand Hydraulic Dynamometer 90 kg[7]. The assessment was performed while the child was sitting upright in a chair without back support, arms resting at the sides of the body, elbows flexed 90°, forearms in a neutral position, wrists 0-30° extended and 0–15° ulnar deviation. Three maximal voluntary isometric contractions of both hands, separately, were required and the mean value was recorded as kilogram/force (kg/f) for each child[12]. Hand grip strength measurement was found to have high test-retest reliability (0.99), and determined to be significantly correlated with the total manual muscle strength related to hand movements[11].

Functional measures

The general functional capacity of children was assessed by using the Turkish version of the EK2. This is an outcome measure developed to evaluate general function in non-ambulatory pediatric neuromuscular diseases such as spinal muscular atrophy and DMD. It evaluates 17 functions such as daytime fatigue, head control, cough, and wheelchair use. Each item is scored between 0 and 3. Higher scores indicate lower global functional capacity. The Turkish version of the EK2 was found to have high internal consistency and intra-class correlation[13].

The performance of upper limbs was measured using the PUL scale, developed by Mayhew et al. in 2013 for children with DMD[14]. The PUL comprises 22 items in total, with 21 items that assess the upper extremity functions at the distal (wrist-hand), mid (elbow) and high (shoulder) levels and one item (entry item) that indicates the general upper extremity functional level. The items are scored on a number system ranging from 0–1 to 0–6. A child can score a maximum of 16 points from the high level, 34 points from

the mid-level, and 24 points from the distal level for a maximum score of 74. It has been shown that the PUL is reliable and can cover the all activities from early signs of proximal involvement in ambulant children to distal involvement in non-ambulant children[15].

The ABILHAND-Kids questionnaire, which has been found to be a valid and reliable method to evaluate the difficulty level of activities requiring manual ability in children with neuromuscular disease[16], was used to assess the distal ability of the DMD children. The questionnaire includes 18 items such as opening a packet of chips and dealing cards. Each item is scored as 0 (impossible), 1 (difficult), 2 (easy), and higher scores indicate good manual ability. Also, the ABILHAND-Kids has excellent responsiveness, reliability and validity in determining the difficulties of children with neuromuscular diseases in hand-related abilities during daily activities[17].

Assessment procedure

A physiotherapist with five years' experience in pediatric neuromuscular physiotherapy performed all assessments. All assessments were conducted within the same session and there were resting intervals during the procedures so as not to cause fatigue. Measurements were systematically applied to each child in the order mentioned above. All measurements lasted a maximum of 90 minutes including resting intervals.

Statistical analysis

The IBM Statistical Package for the Social Sciences (SPSS) Version 20 was used to analyze the data obtained from the assessments. The descriptive statistics of the quantitative data were shown by using minimum, maximum, mean ± standard deviation (X ± SD), median and qualitative data as number (n) and frequency (%). The compliance of the variables with normal distribution was determined by using Skewness-Kurtosis and histogram analysis, the Kolmogorov-Smirnov test, and the variant coefficient. Since the data did not comply with normal distribution, Spearman's correlation coefficient (rho) was used to analyze associations between parameters. The level of significance of the associations was determined as r = 0.70-1.00 strong; r = 0.30-0.70 moderate; r = 0.00-0.30 weak or insignificant. The statistical significance level (p) was accepted as 0.05.

RESULTS

Forty-five children with a DMD diagnosis met the inclusion criteria at the beginning of the study. However, seven children were excluded because of poor motivation during the assessment sessions and inability to grasp the dynamometer. Thirty-eight children practising a physiotherapy program at home, with a regular follow-up program once every six months in a physical therapy unit, were included in the study.

The descriptive characteristics of the 38 children included in the study are given in Table 1. All of the children with DMD in this study were determined to be right-handed.

Grip strength of the dominant hand and results related to general functional capacity, upper extremity performance, and manual ability of the children with DMD are given in Table 2. The non-dominant hand grip strength of children with DMD was 1.29 ± 2.03. The grip strength of the dominant hand was statistically higher than the non-dominant hand ($p < 0.05$).

Hand grip strength was not correlated with the height, weight, body mass index, duration of corticosteroid treatment, and duration of the non-ambulatory phase in this study ($p > 0.05$). The Figure shows the correlation of the dominant hand grip strength with functional measures. A moderate, negative, statistically significant correlation was found between the dominant hand grip strength and the EK2 ($p = 0.02$, $r = -0.50$). Similarly, there was a correlation between the non-dominant hand grip strength and the EK2 ($p = 0.03$, $r = -0.46$).

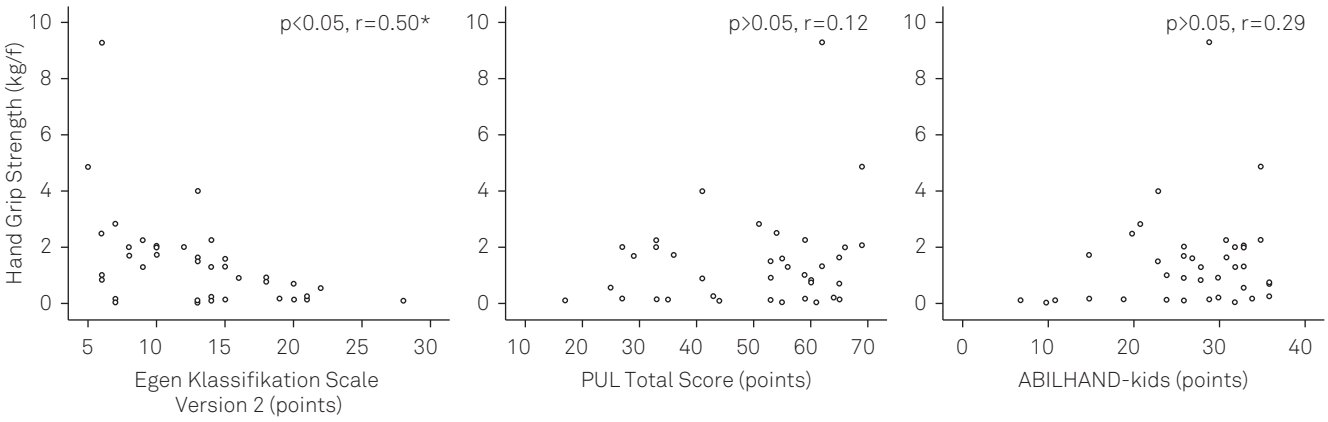
Table 1. The descriptive characteristics of non-ambulant children with DMD included in the study (n = 38).

Parameters	Min	Max	Mean ± SD	Median
Age (years)	8.25	15.83	12.02 ± 1.99	12.00
Height (cm)	125.0	162.0	143.73 ± 10.64	142.50
Weight (kg)	23.10	68.00	42.81 ± 11.13	44.00
Body mass index (kg/cm^2)	13.36	27.18	20.50 ± 3.87	20.21
Duration of corticosteroid treatment (years)	3.04	10.96	6.64 ± 2.06	7.04
Duration of non-ambulatory phase (years)	0.28	6.53	2.41 ± 1.42	2.36

Table 2. Results of hand grip strength and functional measures of children with DMD included in the study (n = 38).

Functional measures	Min	Max	Mean ± SD	Median
Hand Grip Strength* (kg/f)	0.03	9.30	1.45 ± 1.99	1.15
EK 2 (0–51 points)	5.00	28.00	13.02 ± 5.50	13.00
PUL Total Score (0–74 points)	17.00	69.00	49.86 ± 14.34	54.50
ABILHAND-Kids (0–36 points)	7.00	36.00	26.81 ± 7.59	28.50

PUL: Performance of Upper Limb, EK2: Egen Klassifikation Scale Version 2, *Hand grip strength of dominant hand.



PUL: The Performance of Upper Limb. *p < 0.05.** hand grip strength of dominant hand.

Figure. Associations between hand grip strength** and other functional parameters (n = 38).

DISCUSSION

The current study, in which hand grip strength of non-ambulatory DMD children was assessed to determine whether it was associated with global/regional functional measures, indicated a moderate correlation of hand grip strength with the global functional capacity measure—the EK2—in children with DMD who had lost independent walking ability. The results showed that the hand grip strength measurement may be a practical option to quickly estimate global functional capacity of children with DMD in the later phase of the disease in clinical settings, when there is not enough time for a detailed functional assessment by therapists and clinicians.

It has been known that children with DMD produce a lower distal muscle force when compared with healthy peers, even in the early stage of their disease, and the weakness becomes more striking with age[8,11]. The deterioration in manual skills, as well as upper extremity performance, in neuromuscular diseases have also been found to be affected, not only from proximal but also from distal muscle weakness, in two different studies, and a moderate level of correlation between muscular strength and function was proven in ambulatory children with DMD or spinal muscular atrophy[8,18]. The current study showed that hand grip strength of non-ambulatory DMD children was seriously affected when compared with previously-published strength values of healthy peers[19], and the dominant hand of the DMD children was slightly stronger than the non-dominant hand, which corroborates the literature[11]. However, the lack of association between hand grip strength and body mass index, duration of corticosteroid treatment, and duration of non-ambulatory phase may be explained by the variable course of disease for each patient, limited number of participants, and the wide range of descriptive characteristics such as age, height, weight and body mass index of the study population.

The measure of hand strength has previously been reported as being used in pediatric neuromuscular diseases such as spinal muscular atrophy and DMD[7,20]. Dynamometric measurement of gross grip strength, which has previously been determined to be correlated with manual muscle testing, was preferred in this study rather than manually testing many hand muscles, considering the muscles that are prone to fatigue easily in these children, especially in advanced stages of the disease, and its time efficient usage. Although global hand strength was reported to have low validity in early stage myopathy[21], Mattar et al.[11] showed that it was correlated with the Brooke Upper and Vignos Lower Extremity Functional Classification Scales used in the classification of the functional level of upper and lower limbs in DMD. It was also emphasized that hand grip strength could give information about the global hand muscle strength that was evaluated by manual muscle testing[11]. A small reduction of hand muscle strength was reported to be accompanied by a large reduction in timed functional tests (running nine meters and getting up from the floor) in another study that evaluated proximal and distal muscles[8]. The findings of our study suggest that the measurement of hand grip strength with a dynamometer is also moderately associated with global functional ability, in addition to the above-mentioned studies that encourage the use of this measure when a clinician/therapist needs a quick overview of functional status in limited time conditions.

The reason why the hand grip strength did not correlate with the ABILHAND-Kids may be that the scale evaluates the ability of children to manage daily activities that require upper limb usage. Children with neuromuscular disorders may still be functional in some daily activities by using compensatory mechanisms, even if their isolated hand grip muscle strength is inadequate. Furthermore, the lack of association between grip strength and the PUL may be due to the fact that the high-level items in the PUL were

significantly affected in most of the children included in this study, and items at other levels (mid and distal levels) did not consider hand grip function and could be performed without excessive effort. The limited number of children in the study group, heterogeneous ages, hand grip strength, and differences in functional performance scores may be reasons for the lack of relationship between grip strength and the functional performance assessments of the upper limb. In spite of allowing resting intervals between the assessments, the relatively high duration of time required to complete all the assessments performed by the children, can be considered as another limitation of the current study.

In conclusion, although it does not replace a comprehensive functional and strength analysis, a hand grip strength assessment using a dynamometer may be advantageous in cases of time constraints in the clinical settings for clinicians and therapists, in which an immediate decision about global functioning of non-ambulatory children with DMD is needed. Further studies in a large group of children with DMD with homogeneous ages, grip strength, and functional levels are needed.

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