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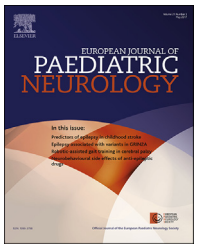

Original article

Neck flexor muscle strength and its relation with functional performance in Duchenne muscular dystrophy

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ABSTRACT

Aim: The aim of this study was to investigate the relationship between neck flexor muscle strength and functional performance in children with DMD.

Methods: A total of 70 children with DMD between Level 1 and 3 according to Brooke Lower Extremity Functional Classification (BLEFC) were included in the study. Children were divided into 2 groups according to neck flexor strength measured by Medical Research Council Scale as Group 1 (3^- and below) and Group 2 (3 and above). Functional performance was assessed by 6 Minute Walk Distance (6MWD) and timed performance tests, and ambulatory status by North Star Ambulatory Assessment (NSAA). Correlations between neck flexor strength and performance tests were analyzed by using Spearman's correlation coefficient in non-parametric conditions.

Results: Thirty-six and 34 children were included in Group 1 and 2, respectively. No statistically significant difference was found in BLEFC between groups ($z = -1.225$, $p > 0.05$). 6MWD ($z = -2.574$, $p = 0.01$) was found to be longer and NSAA ($z = -2.565$, $p = 0.01$) was higher in Group 2. Positive, moderate, statistically significant correlations were determined between neck flexor muscle strength and 6MWD ($p < 0.01$, $r = 0.374$); NSAA ($p < 0.01$, $r = 0.399$) while a negative, weak correlation between neck flexor muscle strength and the duration for standing from supine position ($p = 0.02$, $r = -0.290$).

Conclusions: The results indicate the relationship of neck flexor muscle strength and functional performance in DMD children. It is thought that the approaches to maintain neck flexor muscle strength from the early stages of disease process may support functional activities of daily life with less compensation and effort in DMD.

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1. Introduction

Duchenne muscular dystrophy (DMD) is the most common inherited neuromuscular disease that is caused by a mutation in the gene that encodes for the protein *dystrophin*.[1] Dystrophin is an essential protein for skeletal and cardiac muscles which located in sarcolemma and plays an important role in the maintain of membrane integrity during muscle fiber-contraction. The absence of dystrophin damages the muscle cell integrity and results in progressive muscle fiber-degeneration and replacement with fat and connective tissue.[2] The muscle fiber degeneration in DMD patients leads to progressive loss of skeletal, respiratory and cardiac muscles strength and functional abilities.[3] Progressive and irreversible muscle degeneration also affects ambulation and independency in daily activities and thus leads to further decrease in functional performance.[4] Boys with DMD are generally diagnosed between 3 and 5 years old when they have a clear difficulty in daily routine including standing up from the floor, ascending/descending stairs, walking, and running.[2,5] Such difficulties are caused by the progressive weakness of proximal muscles of extremities, initially in large muscles of hip and shoulder and than distal muscles.[5] Pathophysiological abnormalities and muscle weakness than appear in the neck and trunk muscles of the body which results in postural compensations.[6]

Neck flexion is important to start the sitting from the supine position and standing movements. When the neck flexors weaken, head leaves behind during these activities and the movements can not be performed in full pattern but can only be achieved with compensation. Kuwabara et al. reported that standing from supine position movement which should be performed symmetrically can be initially achieved asymmetrically with compensations in early childhood because the muscle strength development can not be completed in that period of life in healthy children.[7] This movement was reported to advance to symmetric as the children grow. The individuals with weak neck flexors complete the standing from supine and sitting movements by turning into prone, coming to quadripedal position, and then using the neck extensors.[8] Neck flexor muscle strength is also closely related to trunk flexion. In a study which aimed to compare the strength of sternocleidomastoideus (SCM) and rectus abdominus muscles in DMD children who could come to sitting position actively and passively, SCM and abdominal muscles of children who could sit actively were reported to be more stronger.[9] Trunk flexion can not be started in DMD children with weak neck flexors or abdominal muscles are more forced because of weak SCM that the head stays behind which causes a biomechanically disadvantage for coming to sitting position. As a result, the weakening in SCM muscle leads to insufficiency in trunk movements and limitation in functions.[9]

In literature, there are many studies which examine the relation between muscular strength and functional performance in patients with DMD.[10–13] The difficulty in standing from supine position and ascending 4 steps are determined to be affected by the weakness of gluteus maximus, triceps brachii, and quadriceps femoris while holding the handrails when ascending 4 steps are found to be related to the weakness of latissimus dorsi in these studies. As no comprehensive study was observed on this subject in literature, we planned to investigate if neck flexor muscle strength has an effect on outcome measures of functional performance in children with DMD.

2. Materials and methods

2.1. Participants

Ethical approval for this study was received from the Non-Invasive Clinical Research Ethics Committee of Hacettepe University with the protocol number GO 15/38. Signed written consent forms were obtained from all subjects and parents included in the study.

A total of 70 children with DMD were included in the study. Functional status of the subjects were classified according to the Brooke Lower Extremity Functional Classification (BLEFC) which was developed in 1981 by Brooke et al. to classify functional status of DMD children from Level 1 (Walks and climbs stairs without assistance) through Level 10 (is confined to bed) with gradually functional worsening.[14] Subjects in Level 1 (Walks and climbs stairs without assistance), Level 2 (Walks and climbs stairs with aid of railing) and Level 3 (Walks and climbs stairs slowly with aid of railing-over 12 s for 4 standard stairs-) included in the study. Inclusion criteria were proven diagnosis of DMD and being on chronic daily treatment with corticosteroids for more than 6 months. Subjects with known severe cognitive and behavioral disorders impairing compliance with the functional tests and injury or operation related to extremities were excluded.

2.2. Assessments

Demographic data of the subjects were recorded including age, height, weight, body mass index, and the use of corticosteroids. The assessments were performed for all children included in the study were as follows.

2.2.1. Muscle strength

Muscle strength measurements were performed by manually and graded from 0 (no contraction) to 5 (gets full resistance-normal strength) according to subjects' ability to act against gravity while resisting strength offered by examiner.[10,15]

The patient was placed on supine position with the neck extended and the head suspended from the bed. The lower thorax was stabilized. The patient was asked to lift head from the bed against gravity and bring his jaw closer to his chest. If the neck flexion range of motion (ROM) was completed, resistance was given in the direction of extension over the patient's forehead. If the patient did not get resistance, his neck flexor manual muscle strength was evaluated as 3; if he got resistance, the strength value changed between 4 or 5 according to the amount of resistance. If the patient was able to complete the half of the movement, it took 3^{-} value. If the patient was unable to complete the full neck flexion against gravity in supine position, he was required to make full flexion while side-lying. The full flexion ROM was recorded as 2, and

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