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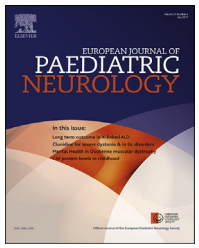

Original article

Effects of functional level on balance in children with Duchenne Muscular Dystrophy

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ABSTRACT

Introduction: This study was planned to compare the static and dynamic balance in children with Duchenne Muscular Dystrophy (DMD) at different functional levels with each other and with healthy peers.

Material and methods: Sixty nine children between the ages of 6 and 11 were included in this study where 52 of them were diagnosed with DMD in Level I (18 patients), Level II (17 patients), and Level III (17 patients) according to Brooke Functional Classification Scale and 17 of them healthy peers were included. In order to assess static and dynamic balance pediatric functional reach test (PFRT) and timed up and go test (TUGT) were used.

Results: When compared in terms of the TUGT, differences were found between all groups, i.e. Level 1 and 2, Level 2 and 3, Level 1 and 3, Healthy peers and Level 1, Healthy peers and Level 2, and Healthy peers and Level 3 ($p < 0.0083$). When compared in terms of the PFRT in the standing positions, there was difference between level 3 and healthy peers ($p < 0.0083$), but not between the other groups.

Conclusions: We found poor functional level in DMD to affect the dynamic and static balance parameters in this study. The dynamic balance of a child with DMD at Level 3 is decreased to a third of a healthy peer.

Duchenne Muscular Dystrophy (DMD) is the most common muscular dystrophy type in childhood. It affects the cardiopulmonary functions and ambulation level and is characterized by progressive muscle weakness.[1,2] The disorder makes up 85% of muscular dystrophies and leads to functional loss due to structural deterioration of the dystrophin protein that provides the connection in the muscle's cell membrane following deletions, duplications or point mutations in the dystrophin gene located in the Xp21.2 region. Symptoms such as duck-like and fingertip walking, pseudohypertrophy of the gastrocnemius and soleus muscles, and Gower's sign (climbing on oneself when trying to stand up) are seen when the child starts walking and at school age.[3] The functional level is maintained between 3 and 6 years in these children. The

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decrease in functional level starts at the age of 6–8 years and functional movements become limited between 9 and 12 years. The person becomes dependent on a wheelchair[4] and cardiopulmonary complications develop, leading to an unfavorable prognosis. These patients generally die in their 20s.[1,4]

Balance is a complex function with many sensory, motor and biomechanical components and both static and dynamic balance need to be preserved to perform and maintain many functional activities.[5] The presence of progressive muscle weakness and contractures deteriorates body balance by affecting the locomotor system in children with DMD, preparing the ground for injuries related to falls. The deterioration of balance in these children can decrease functionality by limiting mobility, independence and social participation due to fear of falls.[6] This has been reported to affect the children physically as well as physiologically.[7] Balance evaluation and training in children with DMD is therefore of great importance.[6]

There is no previous study on the effect of functional level on balance in children with DMD. The aim of our study was to compare the static and dynamic balance in children with DMD at different functional levels with each other and with healthy children.

1. Methods

1.1. Subjects

Children with DMD at the first 3 levels according to the Brooke Lower Extremity Functional Rating Scale and healthy children in a similar age group were included in this cross sectional study. A total of 52 DMD patients with 18 from Level 1, 17 from Level 2 and 17 from Level 3 and 17 healthy individuals were included in the study as a result of statistical power analysis. Name and surname, demographic data and physical characteristics such as height, body weight and body mass index (BMI) of the children with DMD and the healthy children were recorded. All the individuals with DMD consisted of children who have been regularly following-up at our unit, Pediatric Neuromuscular Disorders Unit, in Department of Physiotherapy and Rehabilitation, Faculty of Health Sciences, Hacettepe University, with a home program and family training involving strengthening the proximal muscles of the upper and lower extremities and trunk muscles, stretching the shortened muscles, and performing this program twice a day for 45–60 min taking into consideration of their fatigue levels and have been using an ankle foot orthosis during the night since the last six months.

The children were included in the study after obtaining informed assent from the children with DMD and healthy children and informed consent from their families. Permission was also obtained for the study from Hacettepe University Non-Interventional Clinical Studies Ethics Committee with GO15/279 registration number on 29.04.2015.

The inclusion criteria of the subjects were determined as;

- Diagnosed with Duchenne Muscular Dystrophy and using steroids since the last six months
- Aged 6–11 years,
- Able to cooperate with the instructions of the physiotherapist,
- No acute disease,
- No history of any injury or neurologic or orthopedic surgery within the past 6 months.

Exclusion criteria were the withdrawal of consent and not using steroids.

1.2. Outcome measures

1. Functional level

The functional levels of the subjects were identified according to the Brooke Lower Extremity Functional Classification. This classification method was prepared based on the method determined by Vignos et al.[8] to identify the functional status of the lower and upper extremity during the clinical evaluation of Duchenne Muscular Dystrophy in 1981. The Brooke Lower Extremity Functional Classification scale[9] evaluates the functional level of children with DMD in 10 levels. Level 1 shows the best functional level, level 10 is the most severe level, and the child is dependent on the bed.

Children in the first 3 levels were selected for this study.

Level 1: The child can walk and ascend four stairs without help.

Level 2: The child can walk and ascend four stairs by holding the handrail (walks and ascends the stairs in less than 12 s by holding the handrail).

Level 3: The child can slowly ascend four stairs (walks and ascends the stairs in longer than 12 s by holding the handrail).

2. Balance assessment

In order to assess static and dynamic balance, (a) pediatric functional reach test (PFRT); (b) timed up and go test (TUGT) were used because of clinically easy to use, cheap, and accessible in the clinical setting.[6]

a. Pediatric functional reach test

This test, which is used for assessing static balance, is a reliable and valid modified form of the Function Research Test. It includes subsections for side (left and right) and for forward reaching in the sitting and standing positions.[10] We used forward reaching in the sitting and standing positions which we commonly use in the clinical setting for this study.

Subsections of the test are as follows: sitting in a chair without back support: if the child can sit for 15 s independently, and reach forward in the sitting position; and standing: if the child can stand independently for 15 s and reach forward in the standing position.

The children were asked to lift their arms 90° forward and reach as far as possible while sitting and standing position. Reaching distance was measured in centimeters in each subsection by marking the end point of the third finger over a rule marked on a wall and recorded as "baseline", "final" and "difference"; the total score was obtained by adding the "difference" of forward reaching in the sitting and standing positions.

b. Timed up and go test

The aim of this test, which assesses dynamic balance, is to evaluate balance performance during mobility. It has been