

## Turkish version of the Egen Klassifikation scale version 2: validity and reliability in the Turkish population

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The Egen Klassifikation Scale version 2 (EK2) is an important functional ability assessment scale for nonambulant neuromuscular patients. We investigated the validity and reliability of the EK2 scale in Turkish Duchenne muscular dystrophy (DMD) and spinal muscular atrophy (SMA) patients. Forty-one patients were included in the study. An intra/interobserver reliability study of the EK2 scale as translated into Turkish was performed. The Brooke Functional Classification Scale (BFC) for Upper and Lower Extremities, 10-item Modified Barthel's Index for Activities of Daily Living (MBI), and Pulmonary Dysfunction Index (PDI) were used to investigate the validity of the Turkish version of the EK2 scale. Cronbach's alpha was 0.81 and ICC was 0.87 with a 95% confidence interval ( $r=0.92$ ,  $p<0.01$ ). There were positive, moderate correlations between the EK2 sum score and the BFC upper extremity level and between the EK2 sum score and the PDI, and a negative, moderate correlation between the EK2 sum score and the MBI ( $p\leq 0.01$ ). The Turkish version of the EK2 scale was found to be highly reliable and valid.

**Key words:** neuromuscular diseases, Duchenne muscular dystrophy, spinal muscular atrophy, Egen Klassifikation Scale, validity and reliability.

Neuromuscular diseases (NMDs) are characterized by a heterogeneous group of inherited or acquired disorders of the anterior horn motor neurons, peripheral nerves, neuromuscular junctions or muscles<sup>1-3</sup>. There are 16 different types of inherited NMDs according to the World Muscle Society classification, and many disorders in each NMD subtype<sup>4,5</sup>. Clinical symptoms vary according to the demographic characteristics of the patient, the area of the lesion and the progression of the disease<sup>6</sup>. The general clinical features of NMDs are muscle weakness, premature fatigue during exercise, gait abnormalities, floppiness, delayed motor milestones, sensory loss and pain. These lead to physical limitations in specific daily functions and activities of patients

with NMDs<sup>3,7</sup>.

Duchenne muscular dystrophy (DMD) and spinal muscular atrophy (SMA) are the most frequently seen NMDs of childhood, with general skeletal muscle weakness, which result in serious physical deficiencies<sup>3,8</sup>. Untreated DMD boys usually become wheelchair dependent before their teens, and SMA patients, except for those with type III and IV SMA, spend most of their lives in a nonambulatory state<sup>9,10</sup>.

During the past decade, researchers have conducted many significant clinical trials aimed at treating these diseases, especially DMD. Therefore, clinical outcome measures have been standardized for ambulatory and nonambulatory patients, leading to better

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assessments and results in terms of the efficacy of new drugs<sup>11,12</sup>. Although clinical trials to find a cure for both ambulant and nonambulant patients by targeting the molecular genetic structure of these diseases are ongoing, there is still a lack of sufficient evidence in the literature regarding the effectiveness of the new drugs. Therefore, there is a need for reliable and valid outcome measures for nonambulant patients as well<sup>11, 14, 15</sup>.

The Egen Klassifikation Scale version 2 (EK2) is an important functional ability assessment scale for severely affected patients with neuromuscular disorders. The original version of the Egen Klassifikation Scale (EK) was developed by Steffensen et al.<sup>16</sup> in 2001, largely to meet the need for a detailed functional scale including daily activities for patients in the nonambulatory stages of DMD and SMA. It was updated by adding new categories addressing the need to evaluate daily functioning. EK2 was found to be valid and reliable, and the categories of the EK2 total score were found to be discriminative between both the levels of functional loss and the prognosis for these two genetic disorders<sup>12,16-19</sup>.

The EK scale was originally developed in Danish; two definitive validated translations have been made, in English and Spanish<sup>13</sup>. A Turkish translation and cultural adaptation of the EK2 scale would meet the needs of professionals working in this field in our country.

The purpose of this study was to develop such a cultural adaptation and investigate the validity and intra/interobserver reliability of the resultant Turkish version of the EK2 scale for nonambulatory DMD and SMA patients.

### Materials and Methods

The study was performed in a referral center for the rehabilitation of patients with neuromuscular disorders in Turkey. The necessary written approval was obtained from the creator of the EK scale, Birgit Steffensen of Rehabiliterings Center for Muskelsvind, Denmark. Ethical approval was obtained from the Hacettepe University Noninvasive Clinical Research Ethics Committee, with the protocol number HEK 09/48.

A total of 41 patients who had a genetically confirmed diagnosis of DMD or SMA from a

pediatric neurologist were included in the study. The inclusion criteria were: 1) willingness to participate in the study; 2) age in the range of 2-25; 3) inability to ambulate without assistance; and 4) wheelchair dependency. Patients with severe communication and cooperation problems were excluded from the study. All patients or parents of the patients signed consent forms to participate in the study.

The English version of the EK2 scale was translated into Turkish by two physiotherapists who are experts in their field. The two translations were examined by four expert physiotherapists and converted into a single scale. The final form of the translated scale was back translated into English by an expert who is a native speaker of English and is working in a profession outside of health care. The English translation of the scale and the original form were compared. Necessary adjustments were made after consensus by the committee on the back-translated form. The back translation was presented to the creator of the scale, and after obtaining approval, a pilot study was performed on 10 patients for cultural adaptation. The Turkish version of the EK2 scale was readied for use on patients after adapting the misunderstood terms on the basis of Turkish culture.

Demographic characteristics including the age (in years) and diagnosis of the patients were recorded before performing the assessments. The pretest study of the Turkish version of the EK2 scale was performed on 5 patients and the interobserver and intraobserver reliability studies were performed on all 41 patients afterward. An evaluator used the scale to assess the same patients twice, with a 1-week interval, to investigate intraobserver reliability. Two different evaluators used the scale to assess the same patients separately, with a 1-week interval, to investigate interobserver reliability.

### Characteristics of the EK2 scale

The original EK scale initially contained 10 items and was developed to evaluate the ability to use a wheelchair, to transfer from a wheelchair, to balance in a wheelchair, to stand, to move the arms, to use the arms and hands for eating, to turn in bed, to cough and to speak, and to evaluate physical well-being<sup>16</sup>. In the EK2 version of the scale, 7 more items were added to those just described. The

additional items evaluate daytime fatigue, head control, ability to control a joystick, ability to eat various food textures, ability to eat a meal, ability to swallow and hand function. EK2 presents an ordinal scoring scale from 0 to 3, where the lower scores indicate higher levels of independent functioning and the higher scores indicate lower levels of functioning<sup>16</sup>. The sum of the items ranges between 0 and 51.

The EK2 scale has been shown to be related to muscular strength and pulmonary function as well as valid and reliable for DMD and SMA patients<sup>16,17,20,21</sup>.

The Brooke Functional Classification Scale for Upper and Lower Extremities, the 10-item Modified Barthel's Index for Activities of Daily Living, and the Pulmonary Dysfunction Index were used as gold standards to investigate the validity of the EK2 scale in Turkish DMD and SMA populations.

#### **Brooke Functional Classification Scale for the Upper and Lower Extremities (BFC)**

Brooke et al.<sup>22</sup> were the first to describe BFC as a functional assessment scale for the upper and lower extremities in DMD patients. Vignos et al.<sup>23</sup> had much earlier produced a scale for the lower extremities only. The BFC Lower Extremity Scale classifies the functional status of DMD children from 1 (walks and climbs stairs without assistance) to 10 (confined to bed). The BFC Upper Extremity Scale designates 6 levels of functioning of the arms and hands: 1 (starting with arms at the sides, the patient can abduct the arms in a full circle until they touch above the head and can place a weight of 0.5 kg or more on a shelf above eye level) to 6 (cannot raise hands to the mouth and has no useful functioning of hands)<sup>22,24</sup>. In our study, the evaluator determined functional levels according to the performance of the patients on the BFC Scale for the Upper and Lower Extremities.

#### **10-item Modified Barthel's Index for Activities of Daily Living (MBI)**

The Barthel Index (BI) was first described by Mahoney et al.<sup>25</sup> as an assessment of performance in the activities of daily living. Shah et al.<sup>26</sup> improved the sensitivity and discriminative power of the scale, developing the MBI, which consisted of 10 items testing independency levels in relation to bowel control, bladder control, grooming, toilet use, feeding, transfer (from bed to chair and back), mobility, dressing, stairs and bathing<sup>27</sup>. Performance in these activities is scored 0-3, 0-2, or 0-1. The total score ranges between 0 and 20, where the lowest score indicates a lower independency level and the highest score indicates a higher independency level in activities of daily living (ADLs)<sup>26,27</sup>. In the present study, the same evaluator asked for responses from the patients or their caregivers regarding the patient's performance level in the activities mentioned above.

#### **Pulmonary Dysfunction Index (PDI)**

The PDI was developed in 1988 for clinical use in assessing pulmonary function in patients with expiratory muscle weakness<sup>29,30</sup>. It is accepted as a valid and reliable pulmonary function test for patients with neurological disorders<sup>30</sup>. The index includes four items. Two of the items have to do with coughing strength and difficulty in mucus clearance as self-assessed by the patient. The other two items concern coughing strength and maximum inspiration effort as assessed and scored by the evaluators<sup>28,29,31</sup>. The sum of the scores ranges between 4 and 11, with lower scores indicating fewer clinical signs of pulmonary functional disturbance<sup>31</sup>. For the children included in this study, the same evaluator assessed the first two items with the assistance of the caregivers.

#### **Statistical Analysis**

Statistical analysis was carried out using the Windows-based Statistical Package for the Social Sciences (SPSS) version 18<sup>32,33</sup>. Arithmetical means and standard deviations ( $X \pm SD$ ) for variables determined by measurement and

Table I. Mean Values of the Assessment Tools

Assessment tools	N	min	max	X±SD
Egen Klassifikation Scale Version 2 (0-51 points)	41	3	32	15.41±8.03
10-item Modified Barthel Index (0-20 points)	41	3	14	9.00±2.63
Pulmonary Dysfunction Index (4-11 points)	41	4	11	7.39±1.53

percentages (%) for variables determined by counting were calculated.

Intraclass correlation coefficient (ICC) values with 95% confidence intervals and Spearman correlation analyses were calculated to evaluate intraobserver and interobserver reliability. The intraobserver reliability of the EK2 scale was assessed based on the EK2 sum scores in 8 patients, and the interobserver reliability was assessed based on the EK2 sum scores in 33 patients, who were tested twice at an interval of one week.

Cronbach's alpha values were calculated based on the items. Weighted kappa values were calculated to determine the interobserver agreement based on the items.

The Wilcoxon signed-rank test was applied in nonparametric test conditions based on the items and the EK2 sum score.

The validity of the EK2 scale in the Turkish neuromuscular patient population was determined by assessing the correlations between the EK2 sum score and the BFC upper/lower extremity levels, the MBI and the PDI. The Spearman correlation coefficient was used in nonparametric test conditions. On the basis of the Spearman correlation coefficient (r), the relevance levels of the

correlations were accepted as  $r > 0.70$  = strong;  $0.30 < r < 0.70$  = moderate and  $r < 0.30$  = weak correlation. We considered a significance level of 5% to be important.

## Results

Forty-one patients with the diagnosis of DMD (68.2%; N=28) and SMA (31.7%; N=13), whose mean age was  $13.67 \pm 4.23$  (min: 6.0; max: 24) years old, were assessed.

The mean scores of the EK2 scale, the MBI and the PDI are shown in Table I.

The upper and lower extremity functional levels determined by the BFC Scale are shown in Table II.

### Cultural Adaptation

The EK2 scale was easily adapted to the Turkish neuromuscular patient population. In the pilot study, no items were misunderstood. Completion of all 17 items took 15 minutes. Patients declared that the items were easily understandable and related to daily problems of their children. The back translation of the EK2 scale was approved by the scale's original developer, Birgit Steffensen.

### Reliability

The internal consistency that the evaluator

**Table II.** Distribution of Patients According to the Brooke Functional Classification Scale for Upper/Lower Extremities

	N	%
BFC Upper extremities (1-6 levels)		
Level 1	4	9.8
Level 2	13	31.7
Level 3	10	24.4
Level 4	7	17.1
Level 5	1	2.4
Level 6	6	14.6
Total	41	100
BFC Lower extremities (1-10 levels)		
Level 6	3	7.3
Level 8	6	14.6
Level 9	32	78.0
Total	41	100

obtained from his/her first administration of the scale was found to be high, with a Cronbach's alpha value of 0.81 (N=41). Cronbach's alpha values when each item is deleted individually were calculated and are shown in Table III.

There was no statistically significant difference between the first and second assessments of the same evaluator based on the items (N=8) (p>0.05).

The ICC determined for interobserver reliability was 0.87 with 95% confidence interval (0.7623 to 0.9356); the Spearman correlation coefficient was statistically significant (r=0.92, p<0.01). A mean 1.15±3.58 point difference was found between the sum scores (N=33) obtained from the first and second administration of EK2 by different evaluators; this was not statistically significant (z= -1.944; p>0.05).

Weighted kappa (κ) values of individual items ranged from 0.41 to 0.84 (Table IV). The weighted kappa value for item 14 was not amenable to calculation because all of the patients gave the same answer.

There was no statistically significant difference between the first and second assessments by

**Table III.** Cronbach's Alpha Values with Items Deleted from the EK2 Scale (N=41)

Items	Cronbach's alpha if item deleted
Item 1	0.786
Item 2	0.802
Item 3	0.810
Item 4	0.803
Item 5	0.788
Item 6	0.792
Item 7	0.797
Item 8	0.806
Item 9	0.814
Item 10	0.819
Item 11	0.831
Item 12	0.800
Item 13	0.814
Item 14	0.814
Item 15	0.802
Item 16	0.818
Item 17	0.798

different evaluators for any items (p>0.05).

**Validity**

Forty-one patients were assessed to determine the validity of the Turkish version of the EK2 scale. The correlations between the EK2 sum score and the BFC upper/lower extremity levels, MBI and PDI were assessed in order to study the validity of the scale. There was a positive, moderate, statistically significant correlation between the EK2 sum score and the BFC upper extremity level (p≤0.01) and the PDI (p≤0.01), and a negative, moderate, statistically significant correlation between the EK2 sum score and the MBI (p<0.01). The results of the correlations are given in Table V.

**Discussion**

The EK2 scale includes 17 items targeting functional abilities in essential ADLs of nonambulatory patients with neuromuscular disorders, especially patients with DMD and

**Table IV.** Weighted Kappa Values For Individual Items of the EK2 Scale (N=33)

EK2 scale items	κ
Item 1	0.69
Item 2	0.51
Item 3	0.71
Item 4	0.78
Item 5	0.67
Item 6	0.84
Item 7	0.74
Item 8	0.77
Item 9	0.48
Item 10	0.41
Item 11	0.66
Item 12	0.73
Item 13	0.61
Item 14	N/A
Item 15	0.78
Item 16	0.64
Item 17	0.83

N/A: Not Applicable

SMA. The original English version of the EK scale has been found to be valid and reliable<sup>16, 17</sup>. In this study, we investigated the validity and reliability of a cultural adaptation of the EK2 scale in a Turkish neuromuscular patient population. We found the Turkish version of the EK2 scale to be highly reliable and valid in a Turkish DMD and SMA population.

In the pilot study, we observed that the scale took approximately 10-15 minutes to complete, and no items were misunderstood by patients or their caregivers. The EK2 scale was easy to use in clinical practice and representative of wheelchair-dependent DMD and SMA patients' functional levels and needs. There were no ceiling or floor effects, as was also the case in the original validity study of the EK scale<sup>16</sup>.

The EK2 scale was also seen to differentiate the natural prognosis of Turkish DMD and SMA populations; in particular, the scores of items 14, 15 and 16, which assess feeding functions, were scored differently by DMD and SMA populations. This may reflect the different characteristics of the two diseases. SMA patients have problems in eating and swallowing from the very beginning of the disease process. They usually have proximal weakness, resulting in weak head control and respiratory problems after birth. These problems cause failure in eating foods of all kinds of textures and in swallowing properly without aspiration, in combination with oral motor weakness, at very early stages of SMA<sup>34,35</sup>. In contrast, DMD patients preserve swallowing functions and oral motor muscle strength for a long time, even after they become wheelchair dependent. Similarly, there were differences between the two disease groups

in the scoring of item 3, which evaluates the ability to stand. SMA patients usually have an inability to stand (especially SMA type 2) as well as an inability to stand and walk for even a few seconds at any point during their lives. It is almost always difficult to place them in an upright position in physiotherapy sessions or ADLs even when supported at the knees and hips. To maintain an upright position, they must use knee-ankle-foot or hip-knee-ankle-foot orthoses, but even with those adaptations it is still difficult for them to have a functional gait, as proximal muscle weakness in addition to the weight of the orthoses and the force of gravity inhibits movement of the legs. In comparison, DMD patients can maintain an upright position when supported, even after loss of ambulation. Our findings also support what is known about the natural histories of these diseases and the findings of the study published by Steffensen et al.<sup>16</sup> in 2001.

All category scores and the sum scores of the EK scale were found to be determinative of muscle strength, contractures, forced vital capacity (FVC) and duration of wheelchair dependence<sup>16</sup>. In a subsequent study by Steffensen et al.<sup>17</sup> functional ability, muscle strength, forced vital capacity and clinical status in nonambulatory DMD and SMA patients were described. Muscle strength and FVC were found to decrease within 5 years in both DMD and SMA populations. The decrease per year was found to be three times greater in DMD patients than in patients with SMA. This study<sup>17</sup> showed that EK sum scores changed similarly and correlated with annual changes in muscle strength in DMD patients, while EK sum scores did not change and had weak correlation with annual changes in muscle

**Table V.** Correlations Between the EK2 Sum Score and Other Assessment Tools (N=33)

	Egen Klassifikation Scale (Version 2)	
	r	p
Brooke Functional Classification Scale-upper extremity (1-6 levels)	0.471	0.00*
Brooke Functional Classification Scale- lower extremity (1-10 levels)	0.205	0.19**
10-item Modified Barthel Index (0-20 points)	-0.653	0.00*
Pulmonary Dysfunction Index (4-11 points)	0.389	0.01*

\*p≤0.01; \*\*p>0.05

strength in SMA patients. In addition, the authors found a higher incidence of severe cardiorespiratory problems in nonambulatory DMD patients (16/19) than in nonambulatory SMA patients (4/13)<sup>17</sup>.

Our study included a total of 41 patients in the nonambulatory stage of both diseases: 12 SMA patients and 29 DMD patients. We took into consideration the lower number of SMA patients as being a limitation of our study and therefore assessed the functional levels, independency levels and pulmonary functions of these two neuromuscular disease patient populations together. According to BFC upper extremity results, a wide distribution in functional levels of patients was obviously seen, as levels 1- 6 (Table II). Nevertheless, the majority of patients (n=23; 56 %) were clustered in levels 2 (n=13; 31.70 %) and 3 (n=10; 24.39 %). The patients' upper extremity functional levels were still adequate for performing ADLs, which required arm and hand functioning. The moderate correlation between EK2 sum scores and BFC upper extremity levels indicated the discriminative and selective ability of the scale in assessing the level of daily functioning that would be maintained by nonambulatory neuromuscular patients via the use of their upper extremities.

Lyager et al.<sup>20</sup> described using a simple test to measure FVC along with the EK sum score to determine the risk of forthcoming respiratory failure in a DMD population. Brunherotti MA et al.<sup>21</sup> investigated similar correlations between EK and Barthel Index (BI) scores and pulmonary function parameters—principally FVC—in DMD patients. They found that EK and Barthel Index scores were correlated with FVC and other functional respiratory measures<sup>21</sup>. We assessed patients using the MBI and PDI to determine independence and pulmonary function levels of nonambulatory DMD and SMA patients. In support of the findings of other studies, we concluded that the higher the level of independence in ADLs and pulmonary function in nonambulatory patients, the higher the level of functioning (i.e., the lower the sum score) on the EK2 scale in Turkish DMD and SMA populations.

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