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Reliability of the walking energy cost test and the six-minute walk test in boys with Duchenne muscular dystrophy

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Abstract

The walking energy cost test (WECT) is a useful tool when measuring ambulatory function in children with motor disorders. However, data on the reliability of this test in Duchenne muscular dystrophy (DMD) is not available. In this study we established the reliability of the WECT and the commonly used six-minute walk test (6MWT) in 19 boys with DMD, aged 6–12 years. Participants performed the WECT and 6MWT twice within three weeks. Reliability was determined for walking distance (D, m) and gross energy cost (EC, J kg^{-1} m^{-1}), using the intraclass correlation coefficient ($ICC_{2,1}$) and smallest detectable change (SDC).

Reliability for walking distance was good, with an ICC of 0.92 [95% CI: 0.81–0.97] and 0.83 [CI: 0.53–0.94] for the 6MWT and WECT, respectively, and an ICC of 0.85 [CI: 0.64–0.94] for gross EC. SDCs were 12.2% for D_{6MWT}, 12.7% for D_{WECT} and 18.5% for gross EC. In conclusion, in young boys with DMD, the reliability of both the WECT and 6MWT for assessing walking distance is adequate. Gross EC, as assessed with the WECT is also reliable and sufficiently sensitive to detect change in walking strain following interventions at group level.

Keywords: Duchenne muscular dystrophy; Walking ability; Reliability; Energy cost of walking test; Six minute walking test

1. Introduction

Duchenne muscular dystrophy (DMD) is an X-linked recessive muscle disorder, affecting 1 in 3500 new born boys [1]. The disease is caused by a mutation in the dystrophin gene, which leads to complete loss of dystrophin in muscle cells [2,3]. This results in a muscle wasting that begins in the lower proximal limbs and eventually leads to loss of ambulation and severe disability [4]. The median age of survival is approximately 35 years [5].

Although there is currently no cure for DMD, the increasing availability of promising treatment approaches [6–8] requires the use of well-designed measurement tools in order to provide valid information on the effects of treatments and to monitor disease progression. Clinical observation has clearly demonstrated that walking ability in boys with DMD decreases strongly over time [9–11], indicating that measurement instruments that quantify aspects of walking function are clinically relevant. Such measurement tools should be reliable and sensitive to changes in the population in which the instrument is used.

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One instrument that has been proposed for use in clinical trials in ambulant boys with DMD is the six-minute walk test (6MWT) [12]. This test has been applied in a broad range of therapeutic areas and reflects aspects of normal everyday life, including endurance and walking ability [12–14]. The 6MWT has already been validated in various paediatric populations [15–17], and its relevance to neuromuscular diseases has also been clearly established [18–21], although the use in boys with DMD has only been reported recently [8,12,13,22]. McDonald et al. concluded that the 6MWT offers a clinically meaningful method of outcome assessment in a DMD population, and, also, based on the intraclass correlation coefficient (ICC), they showed that reliability was high (ICC = 0.91 for the walked distance) [12]. However, while these authors reported an encouraging ICC, a full evaluation of reliability requires the reporting of both parameters of reliability and measurement error [23]. Information on measurement error (i.e. precision) is especially relevant when interventions at group level need to be evaluated or when the test is used to determine changes in a specific individual. Therefore, an important limitation in current literature is that evidence regarding the precision of the 6MWT is only available for healthy boys [24].

Another limitation of the 6MWT is that it solely measures walking distance as an aspect of ambulatory function, while two patients walking the same distance may experience a differing physical strain. Increased physical strain (i.e. walking energy cost) is a common and significant problem in children with motor disorders [25–27] and an assessment of the walking energy cost might provide an additional measure of ambulatory function in boys with DMD. However, while studies have provided evidence supporting the reliability of the walking energy cost test (WECT) in children [25,27–29], data on the reliability of this test in DMD patients are not yet available. Moreover, the WECT and 6MWT have never been compared in terms of reliability. Information on the influence of measurement error on statistical power and sample size estimation would provide greater insight when choosing the most suitable test in clinical trials. The aim of our study was to determine the test–retest reliability of the WECT and 6MWT in measuring aspects of ambulatory function in ambulant boys with DMD.

2. Patients

Boys included in the study were recruited from the All Against Duchenne in the Netherlands network. Inclusion criteria were: a confirmed diagnosis of DMD, aged at least 6 years and capable of walking more than 150 m with or without the use of a walking aid. Children had not undergone surgical procedures in the previous six months and had no behavioral problems that would compromise participation in the study. The medical ethics committees of the two participating university hospitals approved the study, and written informed consent was obtained from all participants over 12 years of age or, for younger participants, from their parents.

3. Method

3.1. Procedures

The study was conducted at the departments of Rehabilitation of the VU University Medical Center in Amsterdam and the Radboud University Medical Center in Nijmegen. Each participant visited one of these departments twice over a period of 3 weeks. Test and retest visits were scheduled at the same time of day, and all were performed by the same trained researcher (JK). Two experienced therapists provided training in the 6MWT and WECT. On each visit, a short physical examination was carried out to determine weight, height and leg length, followed by performance of the 6MWT and WECT. To control for influence of fatigue during the experiment, the test order was randomized per child. Furthermore, a 30-min resting period was provided between the two tests (sitting while watching a video).

3.2. Measurements

3.2.1. Walking energy cost test (WECT)

The WECT consisted of a rest test and a walk test. Participants were given specific instructions not to eat or drink for at least 1.5 h prior to testing. Subjects first sat comfortably on a chair for the 6-min rest test (sitting while watching a video), and then performed a 6-min walk test at a self-preferred, comfortable speed on an indoor oval track Testing conditions during the measurements were kept as quiet as possible in order to allow the subject to achieve a steady state. Throughout the WECT, heart rate (HR_{WECT}) was recorded with a polar band (Polar RS400, Polar Electro Oy, Kempele, Finland), and oxygen uptake (VO_2) and carbon dioxide production (VCO_2) were measured with an accurate [30], lightweight gas analysis system (Metamax 3B; Cortex Biophysik, Leipzig, Germany). This system is composed of a facemask, a Triple volume transducer, a gas sample line and a battery-operated unit (650 g) that is worn on the shoulders. After the test, the children's perception of exhaustion (RPE) was scored using the Children's OMNI scale of perceived exertion. This scale uses an indexed category format that contains both pictorial and verbal descriptors positioned along a comparatively narrow numerical response range of 0–10 (with 10 = 'very very tired') [31].

3.2.2. Six minute walk test (6MWT)

The 6MWT consisted of walking for 6 min at fast speed. The test and encouragements were performed according to the method of the American Thoracic Society [32]. The