P.324 - Compensatory trunk movements during functional tasks in patients with Duchenne muscular dystrophy

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interpretation of results. To develop a prognostic model for 1-year change in 6MWD among DMD patients, and to assess the additional predictive value of the model compared to commonly used factors (i.e., age, baseline 6MWD, and steroid use). Natural history data were collected from DMD patients approximately every 6 months over 2 to 5 years. At each visit, patient demographics, treatment experience, and ambulatory outcomes were recorded. Annualized changes in 6MWD were studied between all pairs of visits separated by ~1 year (8–16 months). Prediction models were developed using multivariable regression for repeated measures, and were evaluated using cross-validation. A total of n = 191 ~ 1-year follow-up intervals from n = 39 boys were included. Mean age was 9.4 years and mean 6MWD was 351.8 meters at the start of these intervals; 86.4% had received steroids. The mean annualized change in 6MWD was ~37.0 meters with a standard deviation (SD) of 93.7 meters. Predictions based only on age, baseline 6MWD, and steroid use explained 28% of the variation in annualized 6MWD changes (R-squared = 0.28, residual SD = 79.4 meters). A prognostic model including timed 10-meter walk/run, 4-stair climb, and rise from supine, as well as height and weight, significantly improved prediction, explaining 59% of the variation in annualized 6MWD changes after cross-validation (R-squared = 0.59, residual SD = 59.7 meters). A prognostic model including timed function tests significantly improved prediction of 1-year changes in 6MWD. Explained variation was more than doubled compared to predictions based only on age, baseline 6MWD, and steroid use, indicating significant potential for broader prognostic models to inform clinical trial design and interpretation in DMD.

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Normative data and reference equation for the six-minute walk test in healthy Caucasian boys aged 13–18 years
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Functional capacity assessment in ambulant boys with Duchenne muscular dystrophy (DMD) with the six-minute walk test (6MWT) is relevant to follow-up progression and evaluate treatment efficacy. Due to corticosteroids, the loss of ambulation in boys with DMD has shifted to an older age. Therefore, expanding normative data and reference equation of the 6MWT above the age of 12 is useful. This study aims (1) to collect normative data and develop a reference equation for the 6MWT in typically developing Caucasian boys aged 13–18 years, and (2) to define predictive factors such as anthropometry, leg muscle strength, physical activity and heart rate. A total of 178 typically developing boys (mean age 15 years 5 months, SD 1.45 years) were recruited across five age subcategories between 13 years and 17 years 11 months. The 6MWT was measured as described by Mc Donald et al. During the 6MWT the heart rate was registered with a monitor. Maximal voluntary isometric contractions for knee flexion and extension were recorded with a handheld myometer. Physical activity was evaluated using a standardized questionnaire. The six-minute walk distance (6MWD) increased only slightly with age, from 557 m ± 82 m at 13 years to 577 ± 79 m at 18 years. Age and height related percentile curves of the 6MWD were developed. Weak correlations were found between 6MWD and height (r = 0.15), knee flexion (r = 0.22–0.29), knee extension (r = 0.18–0.19), physical activity (r = 0.15–0.19) and heart rate before the 6MWT (r = 0.19). Comparison of 6MWD with predicted values according to reference equations of Geiger and Ben Saad showed an overestimation by those equations. These insights in the 6MWT and its predictive factors in typically developing boys aged 13–18 years old have to be taken into account when applying this outcome measure in clinical trials in pediatric conditions including boys of this age.

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Duchenne dynamic arm study: Quantitative description of upper extremity function and activity of boys and men with DMD
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Upper extremity (UE) limitations in boys and men with Duchenne Muscular Dystrophy (DMD) have a huge impact on their independence and quality of life. To select interventions that might improve UE function and to assess their effectiveness, in depth insights in the UE are needed. Therefore, this study aims to give a quantitative description of UE function and activity of boys and men with DMD, covering the course of the disease. In addition, the relation between UE functions and UE activity level was explored. Twenty-three DMD patients and 20 healthy controls (7–23 years) participated in this study. Outcome measures on the International Classification of Functioning, Disability and Health (ICF) level of body functions and structures were maximal muscle force, maximal and normalized surface electromyography (sEMG) amplitude, muscle thickness and muscle echogenicity, passive joint angles and joint kinematics during active functional tasks. The Performance of Upper Limb (PUL) scale was used as an outcome at ICF activity level. Different values for subjects versus DMD patients were found for all outcome measures, except muscle thickness. Outcome measures related to proximal UE function could discriminate between patients in different disease stages. Normalized sEMG amplitudes indicated that DMD patients use almost all of their muscle capacity to perform UE tasks. In addition, compensatory muscles become active when the primary muscles reach their maximal capacity. UE functions of DMD patients are already impaired in a very early disease stage and precede the decline in activity level. Active maximal joint angles showed the strongest relation with PUL score (R² = 0.85), followed by maximal muscle force (R² = 0.56), and maximal sEMG amplitude (R² = 0.50). As decline in UE function and activity is already present in an early disease stage, it can be expected that early counteractive interventions will contribute to minimize functional decline in DMD.

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Compensatory trunk movements during functional tasks in patients with Duchenne muscular dystrophy
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Upper extremity function can limit performance of daily activities in boys with Duchenne muscular dystrophy (DMD). When arm function is decreasing, compensatory movements are used to accomplish daily activities. Movement of the trunk is often seen as a compensatory mechanism in DMD. To develop effective interventions or supportive aids it is important to get more insight in the compensatory movements of the trunk in relation to arm function and their relationship with the different disease stages. Therefore, the aim of this study was to investigate the compensatory movements of the trunk in boys and men with DMD and healthy controls while performing functional arm tasks. 23 DMD patients (Brooke 1–5) and 20 healthy controls between 7 and 24 years participated in this study. A 3D motion analysis system (Vicon) was used to record cluster markers on the trunk, while performing a shoulder abduction movement and performance of the upper limb scale (PUL). Four items of the PUL were selected for analysis (reaching, drinking, displacement of a weight and tracing a path), representing performance of various daily activities. Trunk displacement and range of trunk motion in all directions were used to describe the compensatory movements of the trunk. First analyses show larger trunk movement in DMD patients compared to healthy controls. The amplitude of trunk movement increases with disease stage until the patients cannot perform the arm task any more. Boys with Brooke scale 1 already seem to show an increase in trunk movement compared to healthy controls. More detailed
analyses are ongoing. It can already be concluded that the level of arm function and compensatory trunk movements are related in boys with DMD. Thus, it should be kept in mind that the interaction between arm and trunk movements is essential for accomplishing daily task.

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Does neck flexion muscle strength affect function status and performance in children with Duchenne muscular dystrophy?

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The aim of this study was to investigate the effects of neck muscle strength on functional status and performance in boys with Duchenne Muscular Dystrophy (DMD). A total of 70 children with DMD between Levels 1 and 3 according to Brooke Lower Extremity Functional Classification (BLEFC) were included in the study. Neck flexor muscles’ strength was measured by manual muscle testing. The children were divided into 2 groups according to the neck flexor muscle strength as study (neck flexor muscle strength 3 and below) and control (manual neck flexor muscle strength 3 and above) groups. After recording the demographic characteristics, functional performance was assessed by 6 Minute Walk Test (6MWT) and ambulatory status by Northstar Ambulatory Measurement (NSAA). Correlation between neck flexion strength and performance was analyzed with Spearman Correlation Coefficient in non-parametric conditions. Mean age of 36 children (51.4%) in study and 34 (48.6%) children in control groups were 94.3 ± 16.4 and 95.2 ± 20.2 months, respectively. No statistically significant difference was found in functional status measured by BLEFC between groups (z = −1.225, p > 0.05). 6MWT distances were 365.8 ± 75.7 m in study and 405.5 ± 86.4 m in control group while NSAA scores were 19.3 ± 7.5 and 23.6 ± 6.5 in study and control groups. The distance taken in 6MWT (z = −2.574, p = 0.01) was found to be longer and NSAA score (z = −2.565, p = 0.01) was higher in control group. Manual examination of neck muscle strength was determined to give an important clue about functional status and performance of boys with DMD in this study.

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Investigation of the relation between the trunk and upper limb functions in DMD

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The aim of this study was to investigate whether the upper limb functions are affected by trunk control in patients with Duchenne Muscular Dystrophy (DMD). Forty-six children with DMD whose ages were between 6 and 15 years were included in the study. The trunk control of children was determined by Trunk Control Measurement Scale (TCMS). Patients were divided into two groups according to their TCMS score. The children with weak trunk control (0–29 points in TCMS) were included in Group 1 while strong trunk control in Group 2 (30–58 points in TCMS). Upper extremity functions of the groups were assessed with ABILHAND-Kids and The Performance of Upper Limb (PUL). Comparison of upper limb functions at different levels of trunk control was analyzed with Mann–Whitney U Test in non-parametric conditions. The mean age of the children in Groups 1 and 2 were 11.52 ± 2.11 years. The general functional capacity which was assessed by EK2 of DMD children included in the study (p = 0.02, r = 0.47). There is a limited number of functional assessments to determine functional capacity in non-ambulatory children with DMD. This study highlights the importance of hand grip strength – practical and non-tiring assessment in later stage of DMD – as a predictor of general functional capacity in non-ambulatory DMD children.

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Hand grip strength as a predictor of general functional capacity in non-ambulatory children with Duchenne muscular dystrophy

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Determination of functional capacity is very important to guide the rehabilitation of Duchenne muscular dystrophy (DMD). However the number of functional assessments which can be used to determine functional capacity is limited in later phase of this disease. The aim of our study was to determine whether hand grip strength can be a predictor of general functional capacity in patients with DMD. Twenty-five non-ambulatory DMD children were participated in the study. After recording the demographic characteristics of children, functional capacity was evaluated with the Turkish version of Egen Klassifikation Scale Version 2 (EK2) and hand grip strength with Jebsen Hand Dynamometer by taking the average of three attempts in dominant hand. Correlation between the assessments was analyzed with Spearman Correlation Coefficient in non-parametric conditions. The mean age of the children was 11.52 ± 2.11 years. The general functional capacity which was assessed by EK2 was found to be 12.16 ± 5.52 points. The hand grip strength was found to be 1.37 ± 1.98 kg/f. A negative, moderate, statistically significant correlation was found between hand grip strength and functional capacity in non-ambulatory DMD children included in the study (p = 0.02, r = 0.47). There is a limited number of functional assessments to determine functional capacity in non-ambulatory children with DMD. This study highlights the importance of hand grip strength – practical and non-tiring assessment in later stage of DMD – as a predictor of general functional capacity in non-ambulatory DMD children.

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Longitudinal results of magneto-inertial motion analysis in Duchenne muscular dystrophy ambulant patients

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Clinical trials in Duchenne muscular dystrophy (DMD) face challenges with the variability of the most commonly used primary outcome measure, the six-minute walk test (6MWT). The primary objective of our study is the validation of daily life actimetry performed by the magneto-inertial ActiMyo® system as an outcome measure in trials for ambulant DMD children. From the ActiMyo® measurements, it is possible to reconstruct the device trajectory and precisely calculate variables such as stride speed and stride length. Our method to assess variability indicates that a standard deviation of ActiMyo® variables under 4% can be achieved in a 2-week (180 hours of recordings when worn daily) period of home recording. Such a variability is lower than the one of the 6MWT (roughly ±15%) between two visits for the same patient. At baseline, a good correlation was found between the distances of the 6MWT, the median stride speed, the median stride length, and other variables from the ActiMyo® daily life recording. We present here preliminary longitudinal results. Data recorded with ActiMyo® for the six most compliant patients recruited into the randomized placebo controlled part of the SKIP-NMD trial were analyzed over the first 6