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Assistive and adaptive equipment is routinely recommended for boys with DBMD to prevent/delay contractures and prolong ambulation or maintain mobility when ambulation is lost. There are no reports documenting the frequency of use of such equipment. To document the type and age at initiation of various assistive and adaptive devices by patients with DBMD in population based samples from four states. The MDSTARNet is a multistate collaboration that conducts population based surveillance for individuals with DBMD and annual surveys with their primary caregivers. From April 2007 to May 2008, 372 eligible caregivers from four states (AZ, CO, IA and western NY) were identified and 200 (53.7%) completed a telephone interview. The interview included questions about the type of equipment used including night splints, long leg braces, standers, transfer equipment, scooters, manual and motorized chairs. Frequencies among states were compared with χ^2 tests. Statistical significance was set at $p < 0.05$. The caregivers reported use of equipment and mean initiation age as follows: night splints (51%, 7.5 yrs), long leg braces (36%, 8.9 yrs), standers (17%, 9.7 yrs) transfer equipment (58%, 12.5 yrs), scooter (22%, 9.0 yrs), manual wheelchair (72%, 9.5 yrs) and, motorized wheelchair (68%, 11.5 yrs). There were significant differences among states in regards to the use of night splints and scooters. This is the first multi-state survey of equipment use in patients with DBMD and helps document the care patterns for patients with DBMD in various regions across the USA.

doi:10.1016/j.nmd.2011.06.814

P1.55

Assisted bicycle training delays physical deterioration in boys with Duchenne muscular dystrophy: Results of the randomized controlled trial “no use is disuse”

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Physical training could oppose physical deterioration due to disuse in patients with Duchenne muscular dystrophy (DMD). Disuse is not uncommon since patients increasingly have difficulties to move and thus move less. The no use is disuse (NUD) study is the first RCT in patients with DMD that examined whether an assisted low-intensity physical training is beneficial in terms of preservation muscle endurance and functional abilities. Ambulatory and recently wheelchair-dependent boys with DMD were randomly allocated (2:1) to the intervention or the control group. The intervention group received a six-months assisted bicycle training for the legs and arm. The primary study outcomes were the assisted Six-Minute Bicycle Test (a6MBT) and the Motor Function Measure (MFM). A two-way analysis of variance (ANOVA) was used to examine the effect of training on the primary outcomes. Thirty ambulatory or recently wheelchair-dependent boys (mean age 10.5 ± 2.6 year, 60% ambulant and 40% wheelchair) were allocated to the intervention ($n = 17$) or the control ($n = 13$) group. The two groups were comparable at baseline in terms of age and disease severity. All boys, except 1, completed the training. After six months training, the functional abilities of the intervention group were preserved (MFM% at baseline = 64.8 ± 15.0 , and after training = 67.9 ± 14.43), whereas there was a statistically significant decline in the control group (MFM% at baseline = 67.9 ± 13.4 , and after control period = 61.5 ± 13.0) ($p < 0.001$). The same trend was seen for the a6MBT. Analysis of the secondary outcomes (e.g. muscle strength and timed tests) are currently being conducted and will be presented at the conference. This study shows that an assisted low-intensity physical training can delay the physical deteri-

oration as a result of disuse in both ambulant and wheelchair-dependent patients with DMD.

doi:10.1016/j.nmd.2011.06.815

P1.56

Submaximal exercise effects on mdx mouse model

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Duchenne muscular dystrophy is characterized by progressive muscle wasting of skeletal muscles due to dystrophin absence. Membrane instability leads to degeneration/regeneration of muscle fibers and eventual fibrofatty connective tissue replacement. The susceptibility to dystrophin-deficient muscle to damage has led to concerns regarding exercise. Eccentric or maximal effort exercise has not been recommended. Assessments of submaximal exercise will determine effects of concentric and eccentric types of exercises. Four-week-old mdx and control (BL10) mice were assigned to four groups ($n = 5/\text{group}$): non-exercised control (No Treatment; NT), Voluntary Wheel (VW), Horizontal Treadmill (HT), and Downhill Treadmill (DT) for a period of 10 weeks. The efficacy of exercise regimes were assessed using grip strength, rotarod, muscle force, histological parameters and echocardiography.

There was no statistical difference in mean strength between all 4 groups from baseline and endpoint measures. All groups except VW trended towards decreased strength. Systolic heart function significantly improved in VW ($p < .05$) and HT ($p < .01$). Mean weight in VW mice was significantly different from all other groups ($p = .04$). Adjusted weight was significant between all groups. Previous studies have shown that eccentric exercise (downhill treadmill) causes functional deficits in mdx mice. Our study shows that mild submaximal exercise does not result in functional deficits, but instead shows improvements of some endpoints. Specifically, concentric exercise (VW, HT) showed improvements in heart function. Submaximal exercise may have benefits to other organ systems besides muscle strength. Future analysis will provide more insight on the pathology of exercise on dystrophic muscles with histological assessments for inflammation, regeneration and degeneration.

doi:10.1016/j.nmd.2011.06.816

QUALITY OF LIFE: POSTER PRESENTATIONS

P1.57

Neuropsychological profiles in children with Duchenne muscular dystrophy compared to dyslexic population

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Duchenne muscular dystrophy (DMD) is associated with lack of dystrophin, a structural component of the muscular cell membrane. Dystrophin appears to have also a role as signaling molecule and a brain localization. Cognitive and learning deficits are seen in approximately 30–50% of DMD, with a major impairment in verbal domains still largely uncharacterized. We compared neuropsychological and literacy performances in six ambulatory, school-aged DMD patients with 12 age- and sex-matched children with developmental dyslexia (DD).

Among others, the neuropsychological protocol included WISC-III, reading, writing, and comprehension tests, rapid automatic naming (RAN), working memory, and verbal and semantic fluency tests. Only six of 12 children with DD had a history of language delay (LDD). DMD patients and DD controls presented normal IQ values. All DMD were more impaired than controls with no LDD on most verbal measures including phonological working memory. DMD also shared with the selected DD population a low speed in RAN tests and impaired performances on working-memory tests. Preliminary observations confirm that DMD patients are at risk for learning problems with a specific deficit in the speed of lexical access. Difficulties in written language could also stem from a sub-optimal functioning of phonological working-memory. It is helpful to identify such profile in order to offer more appropriate educational measures warranting better performances at school.

doi:10.1016/j.nmd.2011.06.817

P1.58

Parental stress levels in parents of children with muscular dystrophy

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Yoshimura, Sasaki, Akimoto and Yoshimura (1989) suggested that children and adults with conditions, including Duchenne muscular dystrophy and myotonic dystrophy, would particularly benefit from the input of a clinical psychologist to help families develop effective management strategies. Through funding from the Muscular Dystrophy Campaign, a Clinical Psychologist joined the Neuromuscular Team. As well as assessing the effectiveness of psychological intervention through prospective audit; since no baseline information on stress levels in parents of children with muscular dystrophy using the service were available, a research project was developed to address this. Its aim is to determine whether there are differences in the levels of stress experienced by parents whose children have been diagnosed with a specific neuromuscular condition, for example Duchenne muscular dystrophy or congenital myotonic dystrophy; and also whether gender differences exist between parents. This study uses the Paediatric Inventory for Parents (PIP) (Streisand, Branietcki, Tercyak and Kazak, 2001) which measures levels of stress in parents of children with a critical illness. It consists of 42 questions based upon difficult events which parents of children who have (or have had) a serious illness, sometimes face, for example, "Speaking with doctor". Using a 5 point scale parents are asked to indicate (a) how often the event occurred in the past 7 days, and (b) how difficult it was/or generally is for them to cope with. Multilevel analysis will determine whether significant differences exist for (a) condition and (b) gender; and will help guide future delivery of care provision and enable service development.

doi:10.1016/j.nmd.2011.06.818

P1.59

Pre-diagnostic parent experiences in Duchenne and Becker muscular dystrophies, congenital muscular dystrophies, and spinal muscular atrophies: A survey of the national task force for the early identification of childhood neuromuscular disorders

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Describe the pre-diagnostic experiences of parents of children with Duchenne, Becker, congenital muscular dystrophies and spinal muscular atrophy, including parents' descriptions of their earliest concerns. We know little about how parents describe first concerns. Chart reviews indicate that childhood NMDs are often diagnosed well after symptoms emerge, and often well after parents express concerns to a clinician. Cross-sectional, retrospective survey distributed electronically to parents of children diagnosed between ages 6 m–9 y. Data were analyzed from 1087 parents: 651 DMD (60%), 244 SMA (22%), 105 CMD (10%), 87 BMD (8%). Quantitative data largely support published data and clinical experience. Parents reported having concerns before doctors a significant majority of the time. There were no significant differences between those with a positive family history and those without regarding whether parent had concerns before diagnosis. Parents with a family history reported significantly less time between first telling doctor about their worry and age at diagnosis. Parents of first-born children had significantly less time between telling their doctor about their worry and receiving a diagnosis compared with non first-born. Analysis of open-ended questions about words parents use to describe concerns identified themes of milestone-associated concerns, comparing to siblings/peers, and non-specific intuition statements. Concerns clustered in infant/young child (SMA and CMD) and preschool/school age (B/DMD). We report specific words parents use to describe weakness, hypotonia, coordination, muscle hypertrophy, difficulty rising from floor, and others. These data support the previously reported diagnostic delay. Unlike chart reviews, we report parents' words to describe their concerns as one step in facilitating earlier diagnosis.

Funding source: CDC.

doi:10.1016/j.nmd.2011.06.819

P1.60

The effects of psychoanalysis in neuromuscular disorders

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Genetic disorders often affect not only individuals but the entire family. We have hypothesized that a social virus that we have denominated RC (resignation from patients and compassion from relatives) has a negative impact on these families. In progressive neurodegenerative disorders it may also accelerate the progression of the symptoms leading affected patients to exclude themselves from social life prematurely. From September 2006 to August 2010, we have taken 63 subjects belonging to families with affected members into psychoanalytic treatment. Twenty one were non affected relatives and 42 were affected with: 10 myotonic dystrophy, 9 spinocerebellar ataxia, 7 limb-girdle muscular dystrophy, 6 facioscapulohumeral, 2 Charcot Marie Tooth disease, 2 Duchenne, 2 spinal muscular atrophy, 1 Becker, 1 Freidreich ataxia, 1 motor neuron disease and 1 myopathy myotonic proximal. Patients were seen on a weekly basis by a trained team of supervised psychoanalysts. The treatment approach is based on obtaining maximum distance between the subject and the expected feelings that are, hypothetically, caused by his degenerative condition. That is, psychoanalysts did not fulfill the patients' expectation to receive the same amount of compassion that they were used to receiving. Every 3 months the patients were reexamined by the geneticist (Zatz) and by the psychoanalyst (Forbes) who is responsible for supervising the whole team as well as the impact of the treatment reassessed. The results showed that most patients declared that psychoanalysis was beneficial and helped them in dealing in a responsible and inventive way with their disease. Moreover, non-affected relatives resumed their lives by creating a new relationship with the affected relative. We anticipate that psychoanalysis will play a growing role helping individuals dealing not only with genetic disorders but also with the