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Complex biomechanical research methods used for pediatric neurorehabilitation of neuromuscular dystrophy diseases

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CLINICAL ASPECTS OF REHABILITATION

Neuromuscular dystrophy diseases (NMDs) exhibit high degree of heterogeneity, which is generally characterized by decreased muscle strength and muscle fiber integrity. Muscle fibers of different sizes were detected in the dystrophic muscle, as well as the presence of connective tissue elements and fat tissue infiltration could be observed. Twenty-nine different genes have been identified in muscular dystrophy diseases [1]. The flaws of these and unidentified genes cause 34 different genetic disorders that differ both in the severity and onset of the symptoms and in the extent of inheritance. One of the most devastating genetic NMDs is the Duchenne muscular dystrophy (DMD), which is a multilevel/multisystem X-linked disease, that affects 1 in 3600–6000 live male births. The pathological characteristics of DMD include the lack of dystrophin or its isoforms and the subsequent secondary effects on brain development and functioning including social and emotional behaviours as well [2]. The chronic treatment factors (such as glucocorticoid treatment) can also play a part in psychosocial health. Improving the neuromuscular condition of the diseased children requires specific needs that constantly change, sometimes rapidly as the disease progresses.

The basic clinical institution presented the review here is the Neuromusculoskeletal Rehabilitation Center in Budapest at the Szent János Hospital, which also hosts the Neuromuscular Expertise Center for Rare Diseases in Hungary. This institution can be professionally considered as a physical medicine and rehabilitation department for children dealing with neurohabilitation and neurorehabilitation in childhood. It works as a multidisciplinary rehabilitation team as presented in Figure 1. Further preferable professionals may be also included, such as occupational and speech therapists, and special education teachers.

This hospital unit is a patient- and family-centered complex, applying physical, cognitive, and psychosocial elements of therapeutic interventions, i.e., early, preventive neurorehabilitation with movement, sensory, precognitive, and cognitive trainings, furthermore, communication- and learning-centered trainings. From the points of social health care, the profile of the institution is to ensure: (a) from early childhood, the practice of preventive neurorehabilitation for a better quality of life, (b) the prevention of learning disorders, (c) the support in psychosocial integration, and (d) the orientation to a family-centered therapy.

Clinical assessment in DMD is based on standard physical examinations, with the focus on the impairments of the musculoskeletal system and related functional consequences. The regular assessment of the clinical condition is based on measuring muscular strength and range of motion, evaluating posture gait, timed testing of movements, monitoring the ability to scope with activities of daily living, and application of motor function scales. These regular assessments are used to conclude decisions about therapeutic interventions. In the clinical practice, the available objective monitoring methods for rehabilitation needs further development and additional methodological procedures also based on instrumental and technical development. At present, subjective functional scales are dominating functional assessments.

EXPERIMENTAL RESEARCH LINE

Based on priority selection, DMD is in the frontline of this presentation especially because of our present aim that is to

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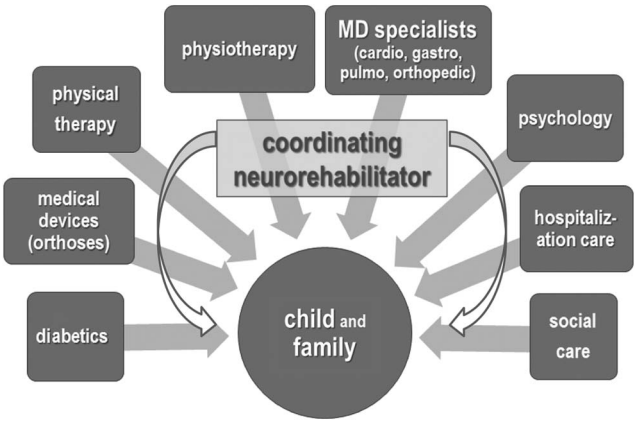

Figure 1. The multidisciplinary rehabilitation team consists of the listed professionals supported by social and hospitalization care and the availability of proper medical devices. The "coordinating neurorehabilitator" physician (at the center of the figure) is a specialist for physical medicine, rehabilitation, and neurology

survey complex biomechanical research methods helping early preventive rehabilitation of this disease. These research methods were developed earlier by our team [3].

To evaluate the functional status of patients with progressive muscular atrophy, first of all in the case of DMD, we need non-invasive instrumental assays, such as stabilometry for posture analysis and to measure coordination abilities, muscle strength assessment, detailed analysis of walking, neuromuscular reaction time, etc., providing objective data for the monitoring of the complex rehabilitation process [4].

METHODS

In our experimental studies using complex biomechanical research methods, the determination of equilibrium and upper-body coordination parameters through registration of pressure-centered trajectories was aimed to carry out for Duchenne (DMD) and Becker muscular dystrophic (BMD) wheelchair-using children. In the presence of parents, 10 muscular dystrophic boys participated in the measurements. Two control groups were created, which were collected from healthy schoolboys of the same age. One control group was examined while sitting in wheelchair and the other without the wheelchair. The average age of examined healthy children was 10.90 ± 3.93 years, whereas the average age of wheelchair-using sick children was 13.67 ± 4.62 years.

The stabilometric parameters were measured in one of the control groups in the standing position, while in case of the DMD and BMD groups and the control group sitting in the wheelchair over a force platform with 1000×1000 mm measuring surface as shown in Figure 2. In addition, a three-channel amplifier, a measuring system containing ADDON microcomputer and laptops, was applied. The Feed 103C software operated the machine (Ing. Büro, Bretz). "Psycho 8" type reaction time measurement system and Dyna 10 universal dynamometer were used to complete the coordination tests.

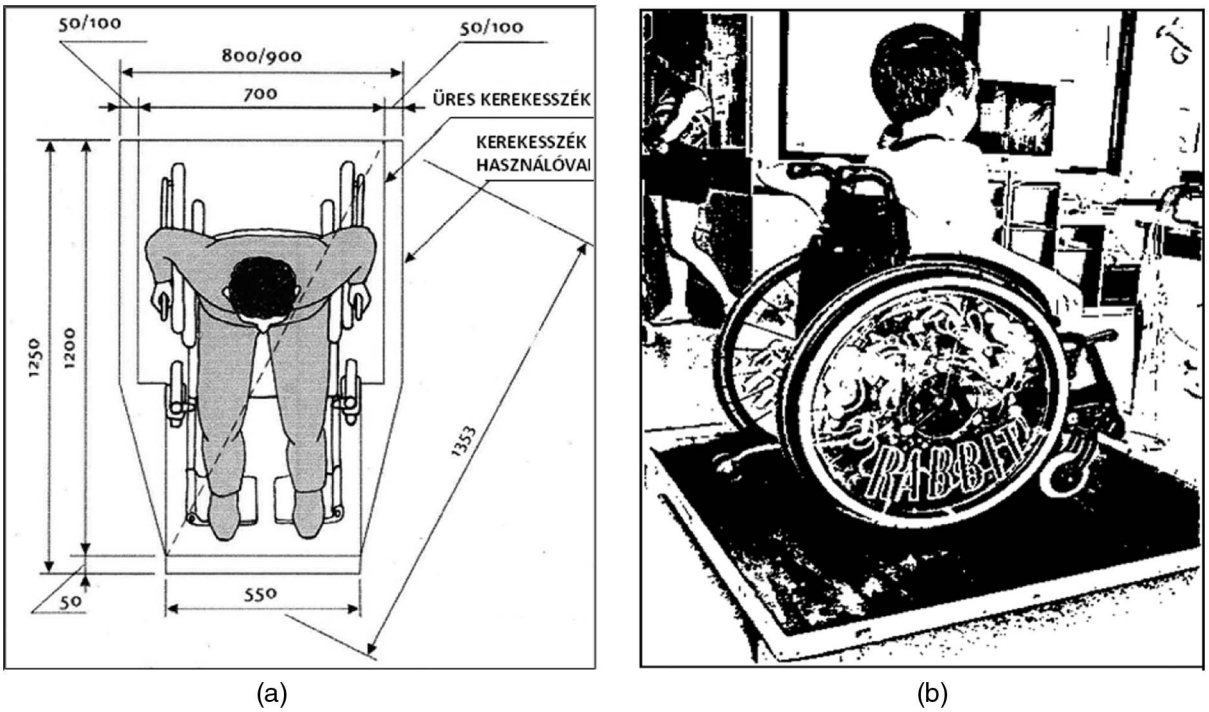

Figure 2. (a) Technical drawing of a wheelchair with standard sizes; (b) stabilometric analysis with a DMD child sitting in a wheelchair positioned on a force platform with 1000×1000 mm measuring surface

Ethics

All procedures were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards (IKEB 555-A/2014).

RESULTS AND CONCLUSIONS

The applied biomechanical functional tests provided informative data on [1] habilitation and rehabilitation of the musculoskeletal condition of DMD children for seat positioning and body posture, on coordinating body posture while sitting in wheelchair, on prevention of asymmetric body posture, preservation of hand function such as choice reaction time, including preservation of combined regulation of neuromuscular and cognitive functions [2]. These novel lines of research are providing important data for rehabilitation in the advanced stage of DMD [3]. The objective data contribute to the selection of more efficient, custom designed orthoses, positioners, and appropriate mobility devices [4]. In addition, gradually achievable postural support may be further emphasized especially for the future studies in helping to reduce the impact of gravity and discourage developing spine statics disturbances leading to asymmetric posture, kyphosis, and scoliosis.

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