

POSSIBILITIES OF COMPLEX REHABILITATION THERAPY IN GENETIC NEUROMUSCULAR AND OSTEOARTICULAR DISORDERS

Cristian Milicin¹, Dan Nemeş^{1,2}, Elena Amăricăi¹, Liliana Caţan¹, George Puenea¹, Daniel Popa^{1,2}

Abstract

Introduction: Genetic neuromuscular and osteoarticular disorders causing a motor and sensory deficit raises not only pathological issues related to its presence, but medical issues in general. Purpose of the study: Highlighting the importance of the complex and sustained medical recovery therapy, installed early in patients with Duchenne muscular dystrophy, Becker muscular dystrophy and Pierre Robin syndrome. **Material and method:** In a period of 5 years, 8 patients with genetic muscular dystrophy were studied, aged between 3 and 13 years, who were evaluated completely and who followed a complex recovery treatment, 3 cures of 15 sessions, with a frequency of 3 sessions per week during one year. **Results:** By applying the complex, sustained and long-term treatment, we obtained a significant improvement of the clinical and functional parameters, in case of all patients studied. **Conclusions:** The complex recovery treatment, applied early, leads to stimulation and acceleration of the nervous regeneration process.

Keywords: Duchenne muscular dystrophy, Becker muscular dystrophy, Pierre Robin syndrome, medical recovery, nervous regeneration.

Introduction

Genetic neuromuscular and osteoarticular disorders determines a motor and sensory deficit, with strong implications on economic, professional, social and not at least, family level.

It raises not only pathology issues related to its presence, but medical issues in general. This pathology causes particular concern of physicians, patients and their entourage, concern about the physical and mental handicap of patients, about the caregivers, and the technical means available for the physician, about the complex treatment possibilities and the appropriate social integration.

The complex therapy of genetic neuromuscular and osteoarticular disorders and the long-term interdisciplinary collaboration are very important, based on the idea that early, complex and sustained therapy, closely monitored by a multi specialized team consisting of: family doctor, neurologist, dentist, medic rehabilitation physician and psychologist, will have a much higher efficiency than individual therapy. (1- Muresanu, 2002)

Objective of the paper

Revealing the importance of early establishment of the medical recovery treatment in patients with Duchenne muscular dystrophy, Becker muscular dystrophy and Pierre Robin syndrome.

Studying the behavior of clinical, functional and electrophysiological parameters in patients with Duchenne muscular dystrophy, Becker muscular dystrophy and Pierre Robin syndrome, before and after the treatment, comparing the results obtained after three sessions of complex medical rehabilitation therapy.

Analysis of the possibilities for stimulating neuronal regeneration, with probable potential of neuroplasticity, after applying the chosen approach.

Material and method

During the period April 2008 – April 2013, at the BFT Clinical Hospital in Timișoara, we studied 8 patients, 3 with Duchene muscular dystrophy, 3 with Becker muscular dystrophy and 2 with Pierre Robin syndrome, aged between 3 and 13 years.

Each patient underwent complex assessment to determine the severity of the disease:

- General assessment: using the FIM (FUNCTIONAL INDEPENDENCE MEASURE) scale (2 – Nemes, 2001)
- Assessment of the muscle strength:
 - Determining the degree of force by manual method according to the method in 6 steps (0-5) of the National Foundation for Infantile Paralysis (2 – Nemes, 2001)
 - Assessment of muscle strength with the pressure gauge, which registers the grip of the patient's affected hand. The results obtained from the affected limb were compared to the results from the healthy limb.
- Assessment of sensitivity: tactile, thermal, painful, vibratory, proprioceptive, kinesthetic.
- Determination of the adjustment coefficient α :
 - The adjustment coefficient α is defined as the ratio between the intensity of triangular current with a duration of 1000 ms and the intensity of rectangular current with the same duration, for values that produce minimal contractions (2 – Nemes, 2001). Determining the adjustment coefficient is very important because its value reflects the degree of denervation of muscles to be subjected to electro-stimulation.

¹Timisoara “Victor Babes” University of Medicine and Pharmacology,

²Timisoara City, Emergency and University Hospital - Rehabilitation and Rheumatology Department

E-mail: cmili27@yahoo.co.uk, nemes.dan@gmail.com, ama.elena@gmail.com,

lilicatan@gmail.com, george.puenea@gmail.com

- Electrodiagnosis was conducted at the level of affected muscles, thus the adjustment coefficient α was determined. (3- Kimura, 2001)

- As a working technique, the bipolar technique for determining curve I/t (normal values for adjustment coefficient α : normo-innervated $\alpha=2.5-6$; partially denervated $\alpha=1-2.5$; totally denervated $\alpha<1$). We used the Siemens Universal – Neuroton 826 device (Figure 1A).

- To determine the adjustment coefficient α we used point electrodes that were placed according to the standard methodology.

• Determination of the motor nervous conduction velocity in the affected muscles: We used the principles and the working method standardized by the northern school (4-Milicic, 2008). We used the Keypoint - EMG/EP SySTEM device manufactured by Alpine Biomed Corp. USA. (Figure 1B)

Each patient received an initial comprehensive assessment, as well as complex assessments before and at the end of each cure of complex treatment and rehabilitation. The main objectives of the physiokinetotherapy treatment applied to the 8 patients were as follows:

1. Prevention and correction of deviations with the help of fixed orthoses and passive mobilizations (Figure 2) (5-Stalberg, 1993).
2. Prevention of joint stiffness: passive mobilizations (Figure 3A), auto-passive, passive-active (Figure 3B), active (Figure 3C), and hidrokinetotherapy.
3. Rare vasculotropic disorders were eliminated using antideclive positions, massage for facilitating the circulation (Figure 4) and elastic gloves.

4. The reeducation of paralyzed muscles was achieved by:

- Electro-stimulation of partially denervated muscles
- Initially: elements of facilitation, contra-lateral exercises, PNFs, Kabat diagonals (D1E, D2F), analytical exercises
- Later we added exercises for increasing muscle strength and resistance (Figure 6).
- Finally, we applied stability and coordination exercises (Figure 7).

5. Restoration of sensitivity (6 – Leonard, 1998), (7- Xharadez, 2002).

Results and discussion

The overall evolution during the 3 sessions was generally good. Comparing the FIM results, initially, with the intermediary ones (at the end of each cure) and with the final ones (after 1 from the beginning of the treatment) they significantly improved (Figure 8).

Most patients reached 3 points on the muscular strength scale determined manually after the 3 cures (Figure 9A) and presented a growth by 0.4-0.6 of the values of the muscular strength measured with the pressure gauge after the three cures (Figure 9B).

There was an increase by 7-9 of the sensitivity assessment values at the level of the affected limb at the end of the treatment in most cases (Figure 9A).

After the treatment, most patients presented a growth by 0.5-0.8 of the values of the adjustment coefficient α (Figure 9A).



A

Figure 1A. Siemens Universal – Neuroton 826 device.



B

Figure 1B. Keypoint - EMG/EP SySTEM.



Figure 2. Fixed orthose.



A

B

C

Figure 3A. Passive mobilizations, Figure 3B. Auto passive-active mobilizations, Figure 3C. Active mobilizations



Figure 4. Massage for facilitating the circulation.



Figure 5. Electro-stimulation of partially denervated muscles.



Figure 6. Exercises for increasing muscle strength and resistance.



Figure 7. Stability and coordination exercises.

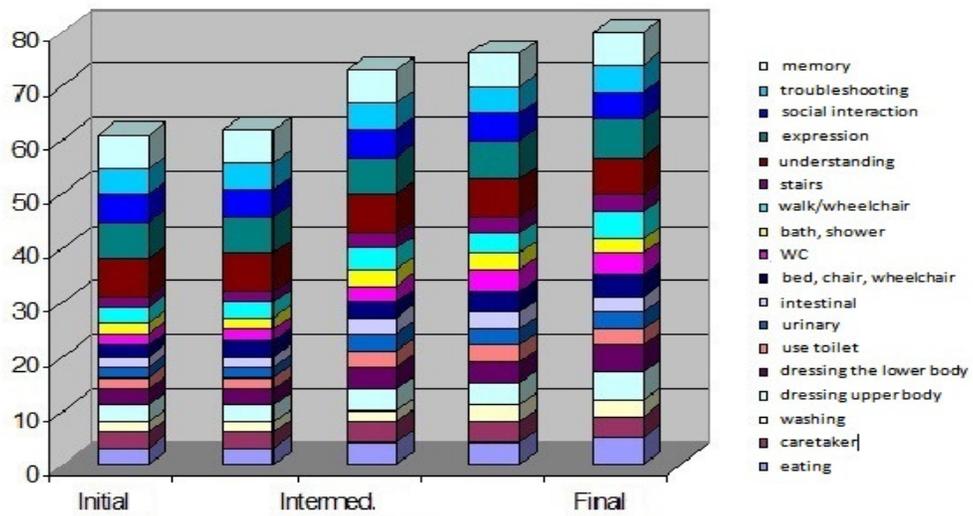


Figure 8. FIM results.

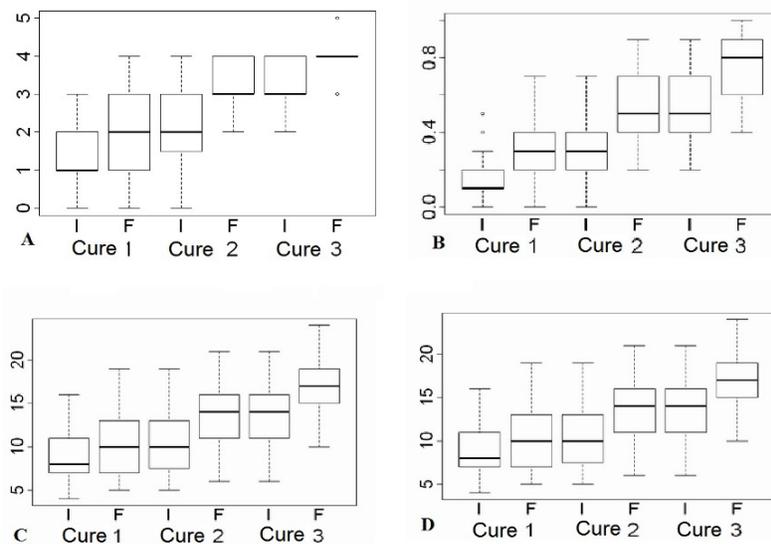


Figure 9. The sensitivity assessment values.

Conclusions

An aggressive approach of the patient with genetic neuromuscular and osteoarticular disorders is essential, but the effort during exercise (regardless of the means of performance) must be progressive, individualized, applied with care, because the increased functional requirements can damage the muscle (as final element in the performance of the movement in the ‘neuro-muscular-artro-kinetic apparatus’) immature or dystrophic, which will not be able to support any over tension (‘overload’).

The clinical and electrodiagnostic assessment of genetic peripheral motor neuron lesions is mandatory to establish the location and the level of nervous damage, as well as a proper therapeutic strategy.

The parameters considered for the study can very well monitor initial deficits and the evolution under treatment.

The stimulation means used were very efficient in the clinical and functional evolution of the genetic peripheral motor neuron syndrome, both for functional and for organic lesions.

The analysis of data from the study allowed us to conclude that the proposed treatment can have two effects for genetic peripheral neuron lesions:

- Stimulation of the nerve by producing supramaximal impulses triggering a motor response
- Stimulation of limb regeneration by accelerating the recovery process of tissues by activating the metabolism and stimulating the cytogenesis.

Taking in consideration the good results obtained in a short time (12 months), we may assume that the used rehabilitation methods also have a neuroplasticity effect.

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Correspondence to:

Cristian Milicin
Timisoara City, Emergency and University Hospital -
Rehabilitation and Rheumatology Department
E-mail: cmili27@yahoo.co.uk