

Comparison of motor function in patients with Duchenne muscular dystrophy in physical therapy in and out of water: 2-year follow-up

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ABSTRACT

The treatment for patients with Duchenne Muscular Dystrophy (DMD) is multidisciplinary. It is necessary to understand the effects of activities performed on dry ground as well as water-immersed to allow the development of intervention protocols. **Objective:** To compare the motor function of children with DMD in physiotherapy carried out on the ground and immersed over a 2-year period. **Method:** A retrospective study, assisted by the Brazilian Association of Muscular Dystrophy (ABDIM), of 23 patients diagnosed with DMD, 8-24 years of age. We collected assessment data both water-immersed (adaptation to the water, bipedalism, sitting position, cross and longitudinal rotation, swimming, and running) and on the ground (Egan Klassifikation Scale and Vignos Scale) over a 2-year period. **Results:** Analyzing the ratings between semesters during the 2-year period, there were differences in the performance of water-immersed activities ($p < 0.001$) and there was no difference on the Egan Klassifikation Scale ($p < 0.003$) or the Vignos Scale ($p < 0.012$). **Conclusion:** Due to the physical properties of the water, patients either improved or maintained their scores for motor function in water immersion. However, their scores for the Egan Klassifikation and the Vignos scales, which represent motor function on the ground, diminished.

Keywords: Muscular Dystrophies, Immersion, Physical Therapy Modalities

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INTRODUCTION

Duchenne Muscular Dystrophy (DMD) is a progressive and irreversible neuromuscular, X-linked disease caused by deficiency in the production of dystrophin. Its incidence is 1 in every 3,500 live births. Clinical manifestations start around the age of five, with difficulties in climbing stairs and walking, in addition to falling frequently, evolving to hyperlordosis, scoliosis, pseudo-hypertrophy of the calf muscles, and a waddling gait.¹⁻⁹

Physiotherapeutic intervention seeks to improve the functional capacity, delay the progression of the disease with low impact activities, train breathing, promote postural alignment, prevent muscle shortenings, and practice activities to prevent fatigue. Hydrotherapy is a resource that has shown to be appropriate for the treatment of progressive muscular dystrophies.¹⁰⁻¹³ The water physical properties facilitate the voluntary movement, postural readjustment, and the relief from pains with less impact on joints. It is common to find children with DMD who use wheelchairs and yet are able to walk when immersed in water.¹⁴⁻¹⁹

It is necessary, based on the organization of evaluation routines, to demonstrate the effects of the various physiotherapy resources that associate ground and water-immersed activities and thereby allow the development of more appropriate intervention protocols and, in the case of children with DMD, less injurious.²⁰⁻²⁴

OBJECTIVE

The objective of this study was to compare the motor function on the ground and in the water of children with DMD, during physiotherapeutic monitoring, over a period of two years.

METHOD

Case studies

It was carried out retrospective study over a period of two years of data from both hydrotherapy (in immersion) and motor therapy (on the ground) evaluations of 23 patients diagnosed with DMD, confirmed by genetic study (DNA), with a mean age of 15.1 ± 4.2 years, treated at the *Associação Brasileira de Distrofia Muscular-ABDIM* (Brazilian Association for Muscular Dystrophy).

The inclusion criteria were that all the patients in the study should be involved in Motor Physiotherapy, Aquatic Physiotherapy, Respiratory Physiotherapy, medical monitoring, and Occupational Therapy. A Rehabilitation Protocol prevented any patient in the sample from having severe cardiopathy or severe ventilatory restriction that could impede the activities on the ground or those in immersion. No patient was excluded from this sample due to their fulfillment of the Institutional Protocol for semester reevaluation in all the departments.

All the participants had one 40 minute-session of aquatic physiotherapy per week, with an average water temperature of 34 °C, with the physiotherapeutic goal of adapting to the liquid environment, training balance both sitting and standing, functional exercises for upper limbs, lower limbs, and trunk, trunk rotations by the Halliwick method, gait training, backstroke swimming, and relaxation while floating.

The patients also had two 50-minute physiotherapy sessions on dry ground each week. In those sessions they practiced stretching, joint movements, active-free, active-assisted, passive, and supine-positioned exercises, activities sitting and standing, balance training, standing with the help of orthoses, and gait.

The respiratory physiotherapy evaluated the respiratory function to prevent pulmonary complications. It consisted of the application of techniques for the capacity of maximum insufflation and efficacy of coughing, in addition to evaluating every week the patients with and without indication of non-invasive ventilation, promoting a better quality of life.

The medical team acted on cardiopathies, preventing and treating them when already installed. Any ventilatory restriction was monitored along with the respiratory physiotherapy and, when needed, it were performed maneuvers for pulmonary expansion, manually-assisted coughing techniques, and introduction of ventilatory support through a non-invasive ventilatory device, in addition to orientation to family members. In this context, vaccination to prevent influenza and pneumococcal infections was introduced, in addition to aggressive antibiotic therapy.

The occupational therapy team had as its main goal the functional rehabilitation of the upper limbs, focusing on the main functional difficulties either in the performance of activities of daily living (feeding, hygiene, dressing, and

locomotion) or on practical living (studies, leisure, and working, among others). The intervention involved prescribing wheelchairs and postural adaptation systems, and the use of assistive technology such as orthoses for the upper limbs and adaptive devices, seeking to improve performance and independence in the performance of activities.

Procedures

The present study was approved by the Ethics in Research Committee from the *Universidade Federal de São Paulo* (UNIFESP) (São Paulo Federal University).

Evaluation during water immersion

The data from the evaluations were classified according to the way the aquatic activities were performed, with 0: did not perform, 1: performs with the aid of distal ends (hands and feet), and 2: performs independently.

Adaptation to liquid environment: evaluates the independence of the patient in the water. If there are any signs of tension such as grabbing the instructor, having apnea, shoulders outside the water, eyes closed, reluctance in putting the face in the water, the participant will be considered not adapted to liquid (grade 0) and if none of those signs are present, the participant will be considered adapted (grade 2).

Balance while standing with the level of immersion to the seventh cervical vertebra (C7): if balance is maintained in the standing posture (grade 2), otherwise (grade 0).

Balance while standing with the level of immersion to the xiphoid process: if balance is maintained at xiphoid process (grade 2), otherwise (grade 0).

Balance while sitting with the level of immersion to the C7: the physiotherapist created turbulence with the board and if the patient maintained balance (grade 2), otherwise (grade 0).

Balance while sitting with the level of immersion to the xiphoid process: the physiotherapist created turbulence with the board and if the patient maintained balance (grade 2), otherwise (grade 0).

Halliwick method transversal rotation (supine to vertical position): if the patient performed it in an independent fashion (grade 2), otherwise (grade 0).

Halliwick method longitudinal rotation on the horizontal plane (rolling): if the patient performed it in independent fashion (grade 2), with help (grade 1), otherwise (grade 0).

Backstroke swimming: divided into simple progression (moving with wrists or with arm strokes along the body) and simplified swimming (moving using upper and lower limbs simultaneously or alternately). If the patient swam (grade 2), otherwise (grade 0).

Gait. Gait is the repetitive sequence of movements of the lower limbs that moves the body forward, while simultaneously maintaining the stability of the trunk. While deambulation is characterized by locomotion via knee and hip flexion, without maintaining orthostatism.^{20,21} If the patient performs the gait and/or deambulation (grade 2), otherwise (grade 0).

Evaluation on the ground

The *Egen Klassifikation* scale (EK) was used for ground evaluation. According to Martinez,¹⁰ it is a reliable scale to measure the degree of functional impairment in the activities of daily living of patients who can walk as well as those in wheelchairs.

The EK scale, validated for Portuguese, is an instrument especially developed to quantify the degree of functional impairment in activities of daily living. It is a useful method for distinguishing the functional performance of daily activities in patients with DMD and spinal muscular atrophy. It shows great potential to determine the functional impairment and evaluate therapeutic interventions. It is composed of 10 items, the score ranges from zero (higher functionality) to three (lower functionality), with a total of 30 points.¹⁰ In the present study, it was decided not to use the Motor Function Measurement (MFM) for the land-based individual functional evaluation, because the focus of its reliability was tested with various types of Muscular Dystrophies and not specifically with DMD patients.

The Vignos scale was also used. This scale is valid for evaluating the staging of the disease, and it is used in various studies that involve functional activities in patients with muscular dystrophy. The Vignos scale was developed in 1960 by Vignos and Archibald.¹⁹ It is considered a gold standard in the evaluation of staging of DMD, for it is widely used in various studies, being simple and precise. According to Archibald, the scale is composed of phases of evolution of the motor status, graduated from 0 to 10, with 0 (pre-clinical phase) and 10 (confinement to bed, needing help for all activities).

Data analysis

The data were classified as non-parametric. The difference between the evaluations was analyzed every semester, and $p < 0.005$ was considered as significant.

RESULTS

Data from the Vignos scale showed means of 5.48 (sd 2.92), 5.57 (sd 2.78), 5.78 (sd 2.66), and 6.13 (sd 2.42), and medians of 7.0, 7.0, 7.0, and 7.0 in the first, second, third, and fourth semesters, respectively. When comparing evaluations between semesters, a $p < 0.012$ was observed, which showed a worsening in motor function, according to the Vignos scale.

The EK scale showed means of 6.78 (sd 6.28), 7.09 (sd 6.44), 7.09 (sd 6.52), and 7.74 (sd 6.50), and medians of 8.0, 7.0, 7.0, and 9.0 in the first, second, third, and fourth semesters, respectively. When comparing evaluations between semesters, a $p < 0.003$ was observed, which showed a worsening in motor function, according to the EK scale.

The data generated in the Evaluations in Water Immersion showed means of 10.35 (sd 4.56), 11.74 (sd 4.50), 12.70 (sd 4.50), and 13.00 (sd 4.16), and medians of 11.00, 12.00, 13.00, and 14.00 in the first, second, third, and fourth semesters, respectively. When comparing evaluations between semesters, a $p < 0.001$ was observed, which showed maintenance and improvement of motor function in immersion.

DISCUSSION

The results showed an increase in the EK and Vignos scores indicating worsening of the motor function performed on the ground and an increase in the immersion scores showed more independence to perform aquatic activities.

The performance of functional activities, balance control, and gait are made difficult on the ground due to the force of gravity, which demands quite a lot from muscles impaired by muscle weakness, in addition to muscle shortenings and contractures. Depending on the intended activity, the physical properties of the water such as viscosity, density, hydrostatic pressure, and buoyancy facilitate moving and body support.¹³

According to Becker,¹³ aquatic physiotherapy offers a different approach when compared to ground activities in the treatment of patients with neurological dysfunctions. Ruoti¹⁴ associated the buoyancy with recovering the control of fast reciprocal movement patterns, while Amanajas¹¹ described aquatic physiotherapy as an effective option due to the promotion of voluntary movement, acquisition of various postures, improvement of functionality,

and gait training. The study also describes that immersion promotes body support and minimizes biomechanical stress in muscles and joints. In addition, aquatic physiotherapy improves blood circulation, promotes increase of muscle strength, range of motion, muscle relaxation, and pain reduction.

The findings from the evaluation on the ground coincided with the works by Reed¹ and Frezza³ who described progressive and marked worsening with loss of muscle strength, confining patients to the use of a wheelchair. Silva et al.²⁰ also reported worsening in the motor condition, with clinical manifestations that varied over the years. Both studies used the Vignos scale, which was also used in the present study.

Os achados da avaliação em solo coincidem com o trabalho realizado por Reed¹ e Frezza³ que descrevem piora progressiva e marcada por perda da força muscular confinando os pacientes ao uso da cadeira de rodas. Silva et al.²⁰ também relata piora do quadro motor, com manifestações clínicas variadas com o passar dos anos

Analyzing the data from each scale on the ground separately, a difference between the semesters was observed, implying that there was a change in the motor activity due to time and, consequently, to the aging of the patients and evolution of the disease. These results suggest that periods of six months are suitable for demonstrating changes in motor function on the ground, during physiotherapeutic monitoring.

In regard to the objectives and conducts defined in the physiotherapeutic treatment that the patients were receiving at the *Associação Brasileira de Distrofia Muscular* (Brazilian Association of Muscular Dystrophy), such as activities to adapt to a liquid environment, balance training sitting and standing, functional exercises for the upper and lower limbs and for the trunk, trunk rotations from the Halliwick method, gait training, backstroke swimming, and relaxation while floating, stretching, joint mobilization, active-free exercises, and active-assisted and passive exercises in the supine position, agree with the studies by Nicolini et al.,²⁵ Albuquerque et al.,²⁶ Franzini et al.,²⁷ and Hecker et al.²⁸

The present study is fundamentally important for clinical practice, for it demonstrates the importance of aquatic physiotherapy to facilitate motor function and describes an evaluation methodology on the ground and immersed for clinical monitoring. This study shows the relevance of monitoring

the motor function on those two media, since motor behavior evolves differently and, thus, this fact should be considered when making decisions on physiotherapeutic conduct. There are no similar studies in the literature.

CONCLUSION

When comparing the evaluations between four semesters over a period of two years, the maintenance of water-immersed functional activities performed and the worsening of functional water-immersed functional activities is due to the physical properties of the water that, as discussed previously, promote ease of movements. In this way, the patient is capable of continuing to perform water-immersed functional activities that, with the progression of the disease, can no longer be performed on the ground.

More longitudinal studies are necessary to verify whether, in the later phase of DMD, motor function in water immersion is also maintained.

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