Chapter

Physical Exercise as a Tool to Delay the Development Process of Duchenne Muscular Dystrophy

Samuel Alexandre Almeida Honório, Marco Batista, Jorge Santos, João Petrica, Helena Mesquita, João Serrano, Jaime Ribeiro and Júlio Martins

Abstract

Several authors reported that the absence of normal physical activity promotes a faster functional loss of several organs and systems, such as the cardiorespiratory system. It is known that scheduling physical activities and regular exercise for DMD patients, when performed based on a thorough functional evaluation, is fundamental for maintaining the quality of life of these children, as well as other associated resources that should be used, whenever possible. Exercise can help DMD patients to maintain and improve muscular strength for performing activities of daily living (ADL) such as stair climbing, slow the rate of increased weakness or contracture development that can prolong ambulation, maintain enough respiratory capacity and strengthened postural muscles, which can slow the onset of scoliosis. There is a need to pass throughout the message to professionals, staff and families who are in this context or who have children with developmental disabilities that exercise and physical activity are an essential factor for maintaining health and well-being throughout the lifespan. That's what we wish and hope with this chapter.

Keywords: exercise, Duchenne muscular dystrophy, physical activity, movement, life quality

1. Introduction

It is known that there are a growing number of parents looking for ways to improve their children's quality of life because of Duchenne muscular dystrophy's consequences.

As a tool, exercise in aquatic environment allows children to achieve skills that can be difficult on the ground. With this in mind, a literature review was carried out to systematize the pediatric sequelae that can be treated with the benefits of physical activity in Duchenne muscular dystrophy [1–4].

Aquatic activity has been a way of stimulating the development of children and expanding the experiences of healthy, disabled or at-risk children [5]. The premature child and those who are at high risk of neurological injury or developmental delay may already have experienced hydrotherapy as an intensive care intervention. Very young babies participate together with their parents in aquatic programs. Within the exercise techniques, it is hydrotherapy that enables the accomplishment

of activities of greater degree of difficulty, providing patients with psychological benefits. If compared to techniques performed on the ground, hydrotherapy due to the physical principles of water facilitates and improves balance, coordination and posture and gives the patient the feeling of safety. For the child the feeling of safety is found in the arms of the parents and in the contact with the body. This was experienced in the sensation of water environment, before birth, and is now again found in the heat of the water.

2. Exercise for Duchenne muscular dystrophy

Hydrotherapy is a classic form of treatment, used with large varieties of functions [6]. The physical properties of heated water promote a facilitating movement and relief of pain, as well as allowing group work and making therapy pleasant, especially for children, who are often unable to perform certain activities in another environment, if not the aquatic. In this study, the effects of immersion in warm water in children with Duchenne muscular dystrophy were measured using HR, O2 saturation, maximal inspiratory pressure (MIP), maximal expiratory pressure (MEP) and oral temperature (OT) measurements. A total of 20 children with Duchenne muscular dystrophy, male, aged 8 to 15 years, participated in this study.

Regarding the heart rate, a mean decrease of 7.3 bpm was observed between the initial immersion period and the pre-immersion period; between the final immersion period and the beginning of the immersion, there was an average increase of 7 bpm and observed a mean decrease of 0.3 bpm between the pre-immersion and final immersion periods. The oral temperature showed an increase of 0.1° C, during the initial immersion period; in relation to the pre-immersion period, this increase is probably associated with the immersion in warm water added to the physical activity, a decrease of 0.1°C between the final immersion period and the initial immersion period.

The O2 saturation showed a decrease in the values obtained after immersion, where there was a decrease of 2.7 between the initial immersion period and the pre-immersion; a 0.9% increase occurred between the initial and final immersion periods and one increase of 1.8% between the values of the final and pre-immersion periods, which were also considered measures of normal physiological adjustments to physical activities.

At the maximum inspiratory pressure, there was a mean decrease of 8 cm of water between the initial period of immersion and the pre-immersion; this change was considered clinically significant; between the periods of immersion, an average increase of 3.8 cm of water was observed of maximal inspiratory pressure.

At the maximum expiratory pressure in relation to the values obtained between the pre-immersion and initial immersion periods, we obtained an increase of 7.4 cm of water, and subsequently between the immersion and thin periods, there was a drop of 6.8 cm of water. This study showed that hydrotherapy is a therapeutic resource that does not represent an overload for children with Duchenne muscular dystrophy. Another study [7] was aimed to verify the benefits of hydrotherapy in improving gait and balance in patients with mild spastic diplegic type of muscular dystrophy, using a proposed protocol. The objective of this study was to analyze the changes in movements in the lower limbs during gait through a treatment protocol using aquatic rehabilitation, to help the child to achieve better gait independence, for which a case study was used, selecting a male child with gait changes.

The protocol was based on relaxation for 5 minutes: muscle stretching of ankle flexors, ankle dorsiflexors, hip adductors, hip and trunk flexors; passive ankle, plantar and dorsiflexion and circular movements; pelvic girdle dissociation; muscle

strengthening of hip and knee extensors and abductors and bicycle and gait training associated with balance training using a 1-pound ankle support and water turbulence. The patient was submitted to the protocol twice a week, lasting 50 minutes each session, totaling seven sessions.

In the initial evaluation, it was observed that the patient, in his functional activities, acquired all the positions without aid except the standing position and the gait. In the evaluation of the gait after the treatment compared to the initial evaluation, it was verified that there were no changes in gait phases. However, when analyzing the patient in the sagittal plane, it is emphasized that the patient used the support of one hand; there was a moment when he wandered without support, evidencing an improvement in the balance. In this study it can be concluded that the application of the proposed protocol in the hydrotherapeutic rehabilitation of the gait was considered efficient due to the good results obtained and proven in the reevaluation as an increase in balance and gait control.

Other authors [8] evaluated individuals with muscle tone disorders, posture and voluntary movement. These disorders are characterized by the lack of control over the movements, by adaptive modifications of the muscular length, resulting in some cases in bone deformities. Neuromotor involvement of this disease may involve distinct parts of the body, resulting in specific topographical classifications (quadriplegia, hemiplegia and diplegia). A child with DMD with diagnosis of hemiplegia, male, 8 months of age, participated in this study. In the therapy performed, a swimming pool with water between 30 and 32°C degrees was used, twice a week with a duration of 30 minutes in the period of August to December of 2004, totaling 40 sessions.

The hydrotherapy sessions consisted of joint mobilizations, stretches, active exercises, Halliwick concept and Bad Ragaz ring method and neuro-evolutionary Bobath treatment adapted in the water with a duration of 30 minutes.

After the hydrotherapy treatment, the child achieved in the area of personal care the following acquisitions: variability of food textures and use of spoon in food, holding an object against gravity and gains in personal hygiene by acquiring partial brushing of teeth and hair and participation in bathing and dressing. It was noted in the area of mobility the condition of sitting in vehicles and moving within it. He was also able to perform transfers from posture to sitting and transfer from the ground to the bed and vice versa, independence in locomotion in internal and external environments, gain of the distance walked. In the social function area, the child acquired comprehension of word meanings and increased vocabulary, comprising complex sentences, aptly naming objects, concomitantly making use of appropriate gestures and problem solving, resulting in greater interaction with children of his or her age. The analysis of this study can be concluded that the application of hydrotherapy in hemiplegic patients provided a sensorimotor improvement in existing functional skills, as well as in the acquisition of others. Thus, the child was better adapted to its development.

According with this line of investigation, another author [9] sought to correlate fat mass and muscle strength, maximum respiratory pressures and respiratory function in individuals with DMD. We selected 68 subjects with DMD. Muscle strength was assessed through manual tests, maximal respiratory pressures through a vacuum gauge and the Vignos test collected by observation. The fat mass was evaluated by bioimpedance, and BMI was also evaluated. Descriptive statistical analysis and regression model construction were performed. A descriptive analysis of the data was performed, and the subjects were divided by quartiles of age. There was a significant correlation between the dependent values, fat percentage and age. Based on this study, it was concluded that there is a correlation between the percentage of fat and muscle strength, respiratory pressures and respiratory function in subjects with DMD.

Another research [10] studied the correlation between fat mass and age in Duchenne muscular dystrophy. Were selected 68 individuals with ages between 5 and 20 years, with molecular diagnosis of DMD. All were submitted to weight and height measurements and to the body composition analysis test with the use of bioimpedance, in the morning, all on the same day. The results were analyzed by grouping the individuals into quartiles of age and showed a body mass index (BMI) of 21 ± 8 kg/m^2. Thus, it was observed that, with the age and degree of sedentarism imposed by the disease, there was an accumulation of body fat and loss of lean mass. They understand that, in fact, more studies are needed related to the nutritional characteristics of these individuals, in order to better clarify the effects of disease and feeding on the percentage gain and fat mass.

An investigation [11] was conducted to evaluate respiratory muscle strength and peak flow in patients with Duchenne muscular dystrophy undergoing noninvasive ventilation and hydrotherapy. Six volunteers of male gender, aged between 13 and 19, were divided into two groups: control (treated with hydrotherapy) and experimental (treated with hydrotherapy associated with NIV), which were evaluated before and after the 10th and 20th sessions. The results showed a significant difference ($p < 0.05$) when we compared MEP between the control and experimental groups after the 10th ($p = 0.025$) and the 20th ($p = 0.005$) sessions. The study demonstrated that NIV was able to influence an increase in life expectancy, according to the patients' own reports, and that hydrotherapy was a favorable therapy in the improvement of the expiratory musculature in patients with DMD. For this purpose, physical activities were carried out once a week in a pool, with a duration of 30 minutes. The main objective was to maintain and stimulate the patient's respiratory function, which was exercised without the use of a life-saving vest or other type of fluid. The pool height was 110 cm in the shallow part and 115 cm in the deepest part. The water was kept warm at 34°C. The activities were carried out with a group of three children besides the teacher. For warm-up exercises, for 10 minutes, involving movements of the body segments, the following activities were carried out to collect rings at the bottom of the pool, to pass under and over flutuators organized in sequence, to enter and exit the flutuators, to blow balls and fish of floating material and to sink balls. For each of these activities, it was requested to perform inspiration out of the water and exhale with the whole body inside the water. In this way the water exerted pressure against the rib cage, and the inspiration occurred against the resistance. At the end, in the period of 3 to 4 minutes, relaxation was carried out with the student floating in the pool. Six measurements were performed, once a month. The first occurred on July 8, 2001, and the last on December 5, 2001. The values were obtained for respiratory rate per minute and vital capacity. No changes were observed in the value of vital capacity between the first and last evaluations, remaining in 800 cm3. Regarding respiratory rate, a decrease from 29 to 26 cycles per minute was observed. It was observed that there was an increase in the thoracic perimeter in normal inspiration and deep inspiration over the 6 months. There was an increase of 1.5 cm in the thoracic perimeter in the normal inspiration and the values obtained in the thoracic perimeter evaluations. According to the author [12] who studies the muscular attrition associated to DMD starts at the beginning of the second childhood and respiratory muscle weakness leads to a series of events that culminate in respiratory complications that worsen considerably at around 10 to 19 years of age. The respiratory complications presented by DMD patients are due, in part, to muscle weakness and thoracic cavity changes caused by scoliosis that affects the patient with disease progression [13].

In another study [14] authors sought to determine the effects of pool physical exercises on the pulmonary function of the person with Duchenne muscular dystrophy. Physiotherapeutic treatment has proven to be important not only in-patient rehabilitation but also in the prevention of imperative changes in this pathology and in teaching to the family, because better results are expected if the parents cooperate.

Examinations of neurodevelopmental status, locomotor system, functional capacity and respiratory system are therefore important. Only then can the main problems be identified, to collect the necessary information and then structure the treatment objectives, which vary according to each specific case. Thus, several protocols have been introduced in order to document the evolution of neuromuscular diseases. Timed functional tests and specific rating scales were designed to document decreased functional capacity. Developed in 1963, the "Vignos Scale for the Classification of Duchenne Muscular Dystrophy Cases" was elaborated [15] which determines the functional degrees of limbs. Repeated studies were developed to determine the reliability of the functional tests, including the Vignos scale, which demonstrated a high degree of reliability. Manual muscle testing (MMT) and its quantitative muscle testing (QMT) were also applied in order to determine its usefulness and reliability to document the evolution of functional decay.

Quantitative muscle testing (QMT) includes determination of isometric strength and is the most direct method to examine the contractile activity of a particular muscle group. It has the advantage of muscle length, joint angle and speed of being kept constant. This test requires the application of special equipment such as an ergometer.

Tests that examine respiratory function, including forced vital capacity (FVC) and forced expiratory volume (FEV) per second, determined with the help of the vitalograph, provide information about the strength of the respiratory muscles. It may be equally interesting to determine the endurance capacity of the respiratory muscles. For professional evaluation of the examination plan to be used, the child's age, ability to follow instructions, availability of equipment, place of examination (clinic, residence, school), available time and (for research purposes or to serve as a basis for treatment) need to previously assessed.

Currently, some specialized sites have a specific record of evaluation of these neuromuscular pathologies, the "MRC grading muscle strength". This sheet assesses the strength of different muscle groups of the upper and lower limbs and head and trunk but also refers to joint limitations caused by contractures as well as motor skills.

Although these assessments are quite comprehensive and complex, they should be repeated regularly for a correct planning of the treatment, according to the current needs of the individual in question. These needs vary according to the stage of the patient, and their treatment must be adapted.

3. Hydrotherapy on Duchenne muscular dystrophy cases: a summary proposal for intervention

Hydrotherapy with temperatures above 30°C will have a beneficial effect on circulation and will improve the elasticity of connective tissue. Particular attention should be paid to the excessive weariness of the child, which very hot water can cause, since fatigue is harmful.

Muscle strengthening techniques are not indicated, as it is reported that they worsen muscle degradation. It is extremely important to prevent contractures and deformities. In this way the exercises should be performed in the most affected regions of the body such as:

Tibiotarsus and feet—the use of instrumental positions with wedges and in vertical or inclined plane, thus using body weight. When the child is seated, the feet should be supported in a neutral position of the tibiotarsus and without abduction of the hips. The purpose of this care is to prolong verticalization, the use of footwear and the absence of pain in the region.

Knees—usually only needed after loss of gait to prevent flexion by retraction of the hamstrings, allowing prolongation of the verticalization and adoption of a more comfortable sleeping position, manual stretching posture and posture with weights in a sitting position, with the basin in retroversion and the lower limbs aligned.

Ankle—in small children it is possible to do the manual stretching in the ventral decubitus, with the knee in 90° bending. In greater angles the instrumental stance is necessary and in passive mobilization. The use of the ventral decubitus should be recommended whenever possible for sleeping, watching television, reading, etc.

Spine—changes occur after loss of gait. The use of orthoses, which can be the shaped vest, is of controversial interest. During the reducible phase, the manual positions of flank opening in the concavity and the passive traction are used.

Shoulders—tardily after loss of gait, make manual stretches of upper trapezius.

Elbows—also after loss of gait. The aim is not to totally avoid bending but to stagnate the angle at which there is the best lever arm for the weakened flexor muscles. Already the supination deficit must be combated, with mobilization and manual postures.

It is also not a goal to completely combat the retraction of the flexors and extensors of the wrist, since from a certain point the tenodesis grasp (passive hand grasp and release induced by wrist extension or flexion) may be the only one possible.

Hydrotherapy is also important in terms of respiratory function, as it depends on the efficacy of respiratory muscles, as well as the degree of bronchial obstruction, once is known the hydrostatic pressure factor on the rib cage. Because of the initial deficit of forced expiration and cough efficacy, maintenance of bronchial clearance is particularly important from early stages. Subsequently, the ability to inhale deeply is lost.

This is particularly important if we note that in the lung development process, the number of pulmonary alveoli stabilizes at about 8 years of age and then increases in size to adult size. If deep inspirations are not performed, which are important for this increase, the alveolar growth is not so great, being a factor of aggravation of the restrictive disease and of the thoracic deformity.

Hydrotherapy should be done regularly as it improves the technique of bronchial clearance and acceleration of the expiratory flow, causing an active (possible while walking) or passive expiration, which causes the secretions to be released up to the coughing zone. Previously, the secretions must be humidified with air or with flutuators. Percussions are often traumatic and vibrations alone are not productive.

The amplitude maintenance techniques are initially activated and then performed using ventilatory assist devices. When vital capacity equals tidal volume, measures may be taken to establish permanent ventilation.

Until a few years ago, physical treatment of DMD was aimed at preserving and stimulating mobility and motility (as far as possible) through "corrective gymnastics, swimming, prophylaxis of contractures, combating inactivity and unnecessary bed rest" [5] which consisted of nondrug treatment. However, physical therapy is much more than "corrective gymnastics", "swimming" and "combating inactivity". Physical therapy traces treatment with goal-based conduits.

The goal of physical therapy is to enable the child to gain control over his or her possible movements, balance and general coordination, delay weakness of the pelvic girdle and scapular muscles, correct postural alignment (standing, sitting, lying down or during movements), balancing muscle work, avoid fatigue, develop the contractile force of respiratory muscles and control breathing through the correct use of the diaphragm and prevent early muscle shortening.

To achieve these goals, a playful treatment is proposed to indulge playfulness of these patients, as they are still children and become bored easily. Physiotherapeutic procedures should be adapted to the age range of the child and are mainly aimed at

delaying clinical evolution and preventing secondary complications (contractures and deformities). In some cases, corrective surgeries and orthotics assist in the treatment.

Free active and isometric exercises are proposed. Playing a wooden doll, because the movements are monoarticular, requires the contraction of a muscle or a reduced group of muscles; on quadruped or weight-bearing position, as it strengthens the scapular and pelvic girdle. During the execution of these exercises, one must seek to evaluate the range of motion (ROM) and muscle strength, request isometric contractions during movements and make use of weight segments as resistance for the muscle group exercised.

The specific respiratory exercises include motivation, such as "flower smelling" and "blow the candle", provided that in dorsal decubitus at 45° of inclination, neither the inspiratory reserve volume nor the expiratory reserve volume (VRE), without using the accessory muscles, nor do resistance to expiration. Taking this into account, it's important to highlight that the effects of inspiratory resistance training on respiratory muscle function were investigated. The authors [11] evaluated 11 patients with DMD and facioscapulohumeral muscular dystrophy, after respiratory training, which consisted of 2 sessions of 15 min per day at home for 6 weeks; there was a significant increase in respiratory muscle endurance, positively correlated with vital capacity ($r = 0.84$, $p < 0.05$) and maximal inspiratory pressure ($r = 0.76$, $p < 0.05$). According to the authors, the improvement of respiratory muscle function may delay the installation of respiratory complications in these patients. In another study on respiratory muscle training with patients with DMD and spinal muscular atrophy, it was found that gains in expiratory muscle strength were rapidly lost with the end of treatment. However, the perception of respiratory effort remained for a longer period, which could be associated with a reduction of respiratory symptoms. In a 6-month study of specific inspiratory muscle training in DMD patients in the advanced stages, the authors realized that, even after 6 months of termination of the training protocol, the respiratory benefits remained for a long period of time.

The activities in the therapeutic balls favor the alignment and flexibility of the spine, stimulate the mechanoreceptors and articular proprioceptors and improve tone and muscle strength, coordination and balance.

The use of hydrotherapy, using methods adapted from Halliwick and Bad Ragaz, is a complementary feature to ground kinesiotherapy, in order to improve muscle strength, respiratory capacity and joint amplitudes and avoid muscular shortening.

The causes of orthopedic contractures in neurological patients are immobilization, muscle weakness and spasticity. The literature describes techniques of treatment of contractures passive stretching, continuous passive mobilization, splinting, electrical stimulation, botulinum toxin injections and tenotomies. There is no consensus on the best way to use the techniques of treatment of contractures, whether combined or isolated in series. Stretching of the sural triceps, ilium-psoas and tibialis-ischemia should be stimulated in the early stages. Short, ankle-foot orthosis (AFO) or long knee-ankle-foot orthosis (KOFO) should be worn at night to prevent muscle shortening. For postural alignment, instruct the child not to stay too long in the same position and give the child the highest body awareness possible. Parents should be instructed and trained to continue home treatment and to encourage their children to engage in age-oriented recreational activities that provide balance, strength and coarse motor coordination. One study [16] followed 204 children with DMD for a period of 8.9 years on average at a research center in the United States. It was able to monitor the effects of physical therapy and orthopedic treatment on lower limb contractures (LLC) and the duration of walking ability. MMI contractures were better controlled when patients performed a combination

	Type	Frequency	Intensity	Duration
Flexibility	Passive/active	Daily	Low	3x (10–30 seg)
Resistance exercises	Short walking Hydrotherapy	Variable 1–6 per week	Low	Variable 1–20 minutes
Muscular strength	Isokinetic	Variable 1–5 per week	Low	Variable 1–3 series 5–15 repetitions

Table 1.
General exercises guidelines for patients with DMD [18].

of daily passive stretches, stood and walked for some periods of the day, had a tenotomy of the calcaneus tendon, transferred posterior tibial tendon and applied KAFO-type orthoses. After 2 years of the use of bracing, the calcaneus contractures were identical in those patients who performed and who did not perform surgeries. Near the fourth-year post-bracing, however, patients who did not undergo surgery had more severe contractures. Five to 7 years after the operation and the use of bracing, the management of contractures was still good, especially in those patients who performed posterior tibial tendon transfer.

Knee contractions were controlled 5 to 7 years after the placement of bracing, with or without surgery. Patients who used bracing were able to walk for an average of 13.6 years, and even after they lost the ability to walk with bracing, the use of orthoses allowed these patients to remain in orthostasis for an additional 2 years. Another study [17] also reinforces the prolongation of gait and orthostasis with the aid of KAFO-type orthoses, but there is no clarification as to whether it is possible to prolong gait functionally.

These procedures are just suggestions. It is up to the technician to choose the most suitable resources available to his/her patients. It is important to note that fatigue and myalgia on the day after the physical therapy session indicate that there was an excess in the number of exercises and their repetitions and that the intensity should be decreased and have more time to rest. Therefore, the main objective to be achieved is to improve the quality of life and the functionality of these children. The quality of life of an adult can be improved by increasing their independence. For the child, the improvement of their quality of life implies the action of playing, however, in a functional way.

In the late stages of the life of the DMD patient, the goal is to comfort the patient: treat pain and dyspnoea, provide palliative care, meet the psychosocial and spiritual needs of the patient and family and respect the patient's and family's choices in what examination and treatment (**Table 1**).

4. Conclusions

A literary survey can be observed that the treatment in pediatrics with the use of hydrotherapy is quite effective in the pathologies that were mentioned for the accomplishment of this work. It can be concluded that the treatment of children in aquatic environment has a great value, due to its positive effects.

Neurofunctional intervention has much to do with DMD and DMB, as patients with these conditions cannot be seen only due to their limitations caused by neurological diseases, which need not only motor care. They need to be seen as people who need interdisciplinary action, since complications occur in the orthopedic and cardiorespiratory fields (not counting other needs, such as psychological and nutritional monitoring).

Special tests, which allow for more precise monitoring of the evolution of the disease, and tools that evaluate the quality of life should be used not only in academic circles. However, they exist to facilitate the recording of information, for later publications in professional scope.

As the natural evolution of these pathologies is already known, it justified the increasingly early performance of the physiotherapist. We also have the task of guiding the caregivers of these children (parents, teachers and family) and referring to the occupational therapist, so that the necessary adaptations are made in the homes, schools and bathrooms. In addition, play adaptations and physical activity can improve the social life of these children.

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Conflict of interest

The authors declare no conflict of interests.

Author details

Samuel Alexandre Almeida Honório[1*], Marco Batista[1], Jorge Santos[1], João Petrica[1], Helena Mesquita[2], João Serrano[1], Jaime Ribeiro[3,4] and Júlio Martins[5]

1 Polytechnic Institute of Castelo Branco, SHERU (Sport, Health and Exercise Research Unit), Castelo Branco, Portugal

2 Polytechnic Institute of Castelo Branco, Interdisciplinary Centre of Social Sciences (CICS.NOVA)/SHERU (Sport, Health and Exercise Research Unit), Castelo Branco, Portugal

3 School of Health Sciences and Centre for Innovative Care and Health Technology (CiTechCare), Polytechnic of Leiria, Portugal

4 Research Centre on Didactics and Technology in the Education of Trainers (CIDTFF), University of Aveiro, Portugal

5 University of Beira Interior, CIDESD, Covilhã, Portugal

*Address all correspondence to: samuelhonorio@hotmail.com

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