83 - USE OF NINTENDO WII IN THE REHABILITATION OF DUCHENNE MUSCULAR DYSTROPHY.

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INTRODUCTION

Muscular dystrophy consists of gradual muscle weakness and spontaneous degeneration of skeletal muscle fibers. Duchenne Muscular Dystrophy (DMD) is considered the most common disease among the neuromuscular ones. It occurs in childhood, with prevalence in males. The disease is recessive, related to the X chromosome (Xp21) resulting in the imbalance of dystrophin protein production. Thus, the destruction of muscle cells and increase of the permeability of these cells happens. The calcium levels within the muscle increase and proteolytic enzymes are activated, resulting in cell damage (MOFFAT, ROSEN and SMITH, 2007). DMD is characterized by signs and symptoms that are shown around the age of two or three years, evolving progressively and irreversibly, resulting in muscle weakness, functional deficit, contractures, deformity and decreased respiratory capacity. It affects skeletal muscles and can reach the cardiac muscles and the nervous system (FACHARDO et al, 2004).

According to Cyrulnik et al (2007), the neurodevelopment and psychological-cognition disorder observed in the dystrophy are result of the partial or complete absence of dystrophin in the central nervous system. This absence is found in the postsynaptic neuron terminals of the cortex, hippocampus and cerebellum, which are areas intensely involved with reasoning and learning. Sampaio and Oliveiras (2004) believe in the association between cognitive functions and children's neuropsychological commitment, suggesting early diagnosis and guidance to the parents and school, mainly in the cognitive and psychomotor sphere.

Besides the delayed motor milestones and speech delays, the mental retardation (MR) is a quite frequent aspect amidst boys with DMD, affecting about 30% of them. This prevalence is higher than what was observed in the general population, in which the MR rates are of 1% approximately. The average intelligence quotient (IQ) of patients with DMD is of 85. Generally, the verbal IQ is more affected than the executive IQ. The severity of the MR does not seem to correlate with the intensity of muscle weakness. In addition to the cognitive dysfunction, a higher frequency of psychiatric comorbidities is observed in this disease, such as attention deficit and hyperactivity disorder (ANDERSON et al, 2002).

Another study assessed psychiatric and behavioral aspects of DMD by applying projective psychological tests. There was observed hypochondriac concerns, signs of isolation feelings, self-deprecation, marginalization and insecurity, in addition to symptoms of anxiety, emotional lability and depression (especially in older patients) (ROCCELLA, PACE and GREGORY, 2003). These manifestations cause problems when adapting to the milieu and lower emotional wellbeing, while a strengthened self-esteem brings stability, favoring the adaptability to the environment, to the emotional well-being and resistance to adversity (SCHLINDWEIN-ZANINI et al., 2008).

The diagnosis can be established through family history, clinical, laboratory and genetic findings and electrophysiological and histological exams too. The enzyme values, mainly the CK (creatine kinase), achieve very high levels. The muscle biopsy and DNA analysis confirm the disease (FACHARDO et al., 2004).

In this context, the treatment is rather limited. However, the presence of Physiotherapy and Neuropsychology in the lives of these patients is of vital importance. Therefore, the intervention includes activities directed towards the movement capabilities of the child in order to maximize the function and cognition, enhancing the patient's quality of life (UMPHRED, 2004).

A proposal that has been used in the rehabilitation includes the video games of the Nintendo Wii, launched in December 2006, which requires a large amount of body movements from the player while providing him motivation. People interact with games and everything that involves imagination (SANTOS, 2010 apud BRESCIANI and tale, 2012). This research is justified by the small number of studies regarding Physiotherapeutic Rehabilitation through video games used in Duchenne Muscular Dystrophy cases.

METHOD

This study consists of a descriptive research, with the objective to account, analyze and correlate facts and phenomena. It seeks to discover, as accurately as possible, the frequency at which a phenomenon occurs, its relationship and connection with others, their nature and characteristics. This research also develops into human and social sciences (HART and BERVIAN, 2002).

The present study is a case study, based on a survey about a specific individual, to examine aspects of his life. The study was conducted during the period from September to November of 2012 at the Physiotherapy School Clinic of the University Center - UNIFACVEST, in the city of Lages/SC.

The sample consisted of one male participant of 10 years old, 1,47m of height, weighting 27 kg, with clinical diagnosis of Duchenne Muscular Dystrophy. For the patient's inclusion in the study, his responsible agreed and signed the Informed Consent Form, so he was able to participate in the exercises program through the Video Game called Wii, of the trademark Nintendo®. The games used were Wii Sports and Mario.

The Patient's assessment was performed by 2 motor scales:

- **EDM – MOTOR DEVELOPMENT MOTOR** - which evaluates the fine and global motor skills, balance, body schema, SPATIAL ORGANIZATION, speech/temporal organization and laterality, also determining the age and motor quotient. The data was tabulated in the EDM software by using descriptive analysis to compare the pre-test and post-test results in the following variables: MA (Motor Age), GMA/GMQ(General Motor Age and General Motor Quotient) and NA (Negative Age Correlation) specific to each motor area (ROSA NETO, 2002).

- **GMFM - GROSS MOTOR FUNCTION MEASURE** - that aims to quantify the change that occurs in the motor function, it consists of 88 items that are measured by the observation of the child or adolescent. The results were obtained by means of a simple percentage calculation accomplished by counting the total of the tasks' success (RUSSELL et al., 2002).

Also, two questionnaires - that were created for this research - were used. One was directed at the patient, composed

by ten questions, in order to verify the patient's relation with Physiotherapeutic Rehabilitation through the Nintendo Wii Video Game. The other one, aimed the child's mother, was composed of three questions involving the sequence of the previous one, during and after the treatment with the games of the Video Game Wii.

A neuropsychological interview (with the mother and the patient) and psychological care, including neuropsychological orientation about the patient (for the mother) was carried out.

The total of sessions was fifteen, of one hour each, twice a week. The games of tennis, golf, baseball and bowling were used for the treatment, all of them aiming at mobilization and range of motion of the upper and lower limbs. In addition to these, we used the "Mario game" that emphasizes waist movements.

For the accomplishment of these activities, the patient was positioned in a sitting position, due to the impossibility to remain in the standing position, not requiring support because his trunk stability. The movements were made by following the verbal commands of the staff member and the action of the games. The patient made the movements of the upper limbs through the controller, combining extension, flexion, abduction and adduction. For the lower limbs the controller was adapted to the patient's feet, more specifically the feet soles, with the goal of favoring the knee extension movement and dorsiflexion, which was limited. At the end of the 15th session a reassessment of the patient was made.

The software Microsoft Word (2007) was used for the tabulation of anthropometric measurements and results, given that they were presented in charts and tables.

RESULTS AND DISCUSSION

A male child of 10 years old, 1,47m of height, weight of 27 kg, resident in the city of Lages/SC. was evaluated.

According to the mother, the child presented normal motor activities and DNPM, except for running and jumping, until the age of 5. From this age on, he walked on his tiptoes, falling frequently. By the age of 6, a calf hypertrophy occurred. The clinical diagnosis of Duchenne Muscular Dystrophy was confirmed by muscle biopsy report, issued in March 2010.

The results of the Development Motor Scale (EDM) of Rosa Neto (2002) can be observed in Table 1, emphasizing that the Global Motor Skills and Balance were not evaluated, because of the impossibility of the patient to stand up.

Table 1 - Motor evaluation results using EDM (Rosa Neto, 2002).

VARIABLE	PRE-TEST	POST-TEST
CA - Chronological age	122	124
GMA - General Motor age	68	78
NA - Negative age	-54	-46
IM1 - Fine Motor Skills	108	132
IM4 - Body Schema	96	96
IM5 - Spatial Organization	84	108
IM6 - Temporal Organization	120	132
GMQ - General Motor Quotient	55,7	62,9
QM1 - Fine Motor Skills	88,5	106,5
QM4 - Body Schema	78,7	77,4
QM5 - Spatial Organization	68,9	87,1
QM6 - Temporal Organization	98,4	106,5

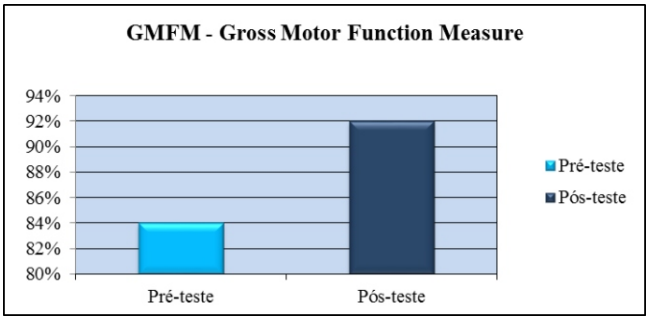
Note: The ages are expressed in months.
Source: data of the research.

In relation to the motor characteristics of the evaluated patient, the Table 1 shows that in 2 months' increase of the CA (122 for 124 months), the patient evolved 10 months of GMA, demonstrating an important evolution. The Negative Age of the pre-test (- 54 months), passed to (- 46 months) in the post-test. That was evident in the General Motor Quotient (GMQ), which rose from 55.7 on pre-test to 62.9 on the post-test, although both have been classified as "substandard". In the DMS, the best performing area was temporal organization (pre and post-test); the worse performance was the spatial organization (on the pre-test) and body schema (post-test).

According to Santos et al (2010), who had evaluated a male child of 7 years old of Chronological Age (CA) and clinical diagnosis of Down Syndrome, the results obtained were 86 months of CA on the pre-test and 90 months on the post test, demonstrating an increase of four months regarding period between the application of the pre-test and the post-test. Regarding the NA (Negative Age), changes on the values were noted, passing from -46 to -42 months in the post-test. The GMA (General Motor Age) increased eight months between the pre-test and post-test, fact that helped on the reduction of NA (Negative Age), positive factor for the Child's development. However the values of the GMQ was classified as “substandard” in both moments (pre-test and post-test).

In the evaluation of the gross motor function, by the GMFM, the difference of values can be visualized in Chart 1.

Chart 1 - Gross motor function data with GMFM.



Source: data of the research.

It was here observed that, through the GMFM scale, both results seen on the pre-test (84%), and on the post-test (92%), had good prognosis, but the child made the post-test with better agility and performance. The D and E stages of the scale were not carried out, due to inability of the patient to stand in orthostatic position by himself.

For Allsop and Ziter (1981), Scott et al (1982) and Brooke et al (1983) apud Silva (2010), who analyzed the motor function of 44 patients with Duchenne Muscular Dystrophy in corticotherapy through GMFM scale in a group of wheelchair users, found that people in this group maintained their motor situation in relation to distal motor skill without significant changes between visits. This is a positive fact, because it demonstrates stabilization of motor function. Moreover, one can observe the decrease of proximal and axial motor loss, as well as stabilization of distal motor function.

With regard to the motivation, some answers of the patient had been: “"I'm satisfied with the rehabilitation through the Nintendo Wii games", "I think the game is fun because it helps in the development of my legs". Answers of the child's mother: "He had no motivation to perform the rehabilitation and began to like it, because it is something different for him, it unites the treatment and amusement at the same time", "the motivation is much higher now."

For Albuquerque and Scalabrin, (2007) apud Sousa (2011), their analysis shows that the virtual environment is a valuable instrument of physiotherapeutic rehabilitation, especially in neurological disorders. It establishes the interaction between patient and the game, what increased the level of motivation of the patient in relation to the treatment and caused the individual to experience a different reality.

CONCLUSION

We can conclude that the use of the Nintendo Wii in the rehabilitation of the patient with Duchenne Muscular Dystrophy increases the motor skills and functional development scores. In the GMFM scale, the patient performed activities with greater agility in post-test. In the EDM, the areas that showed better performances were temporal organization (pre and post-test) and fine motor skill (post-test).

It was evident that the Video Game has increased the level of motivation and patient's satisfaction when performing the treatment, showing interaction with the games.

We also concludes that, due to the progressive muscle degenerative aspect, to the emotional impact and motor and cognitive changes of the disease, it is of great importance the existence of an interdisciplinary therapeutic intervention, which includes Physiotherapy, Psychology, Physical Education, Medicine, among other fields, all together for the benefit of the patient and his family.

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USING THE NINTENDO WII IN REHABILITATION THE DUCHENNE MUSCULAR DYSTROPHY.

ABSTRACT

Introduction: Duchenne Muscular Dystrophy (DMD) is a recessive disease related to the X chromosome. The deficiency of a gene (Xp21) results in imperfection in the production of dystrophin protein. It gradually evolves to muscle weakness. Objectives: to verify the effectiveness of the use of the Nintendo Wii in the rehabilitation of Duchenne Muscular Dystrophy. Methods: A patient with DMD, male, 10 years old participated in the study. The Video Game Nintendo Wii was used for his rehabilitation. The evaluation was carried out using the GMFM (RUSSEL, 2002), EDM (ROSA NETO, 2002), neuropsychological interview and two questionnaires developed for research. Results: Using the GMFM score, it was obtained a good prognosis of 84% on pre-test and 92% on post-test. In the DMS, the best performing area on the pre-test was temporal organization and on the post-test, EDM motor skills and temporal organization; yet, the worse performance on the pre-test was spatial organization and on the post-test, body schema. Through the questionnaires' application, a high level of motivation and satisfaction of the patient was observed. Conclusion: the results showed that the use of the Nintendo Wii in the rehabilitation of the DMD increased motor skills development (temporal organization and fine motor skills) and also reinforced the patient's motivation.

KEYWORDS: Nintendo Wii, Duchenne Muscular Dystrophy, Rehabilitation.

UTILISATION DE LA NINTENDO WII POUR L'AMELIORATION DE LA DYSTROPHIE MUSCULAIRE DE DUCHENNE.

RESUME

Introduction: La dystrophie Musculaire de Duchenne (DMD) est une maladie récessive liée au chromosome X étant donné que la déficience d'un gêne résulte d'un dysfonctionnement de la production des protéines dystrophines. Elle évolue progressivement et provoque de la faiblesse musculaire. Objectifs: vérifier l'efficacité de l'utilisation de la Nintendo Wii sur l'amélioration de la dystrophie musculaire. Méthodes: un patient souffrant de DMD, de sexe masculin, âgée de 10 ans a participé à l'étude. La console Nintendo Wii a été utilisée pour la réhabilitation. L'évaluation a été réalisée grâce au GMFM (RUSSEL, 2002), l'EDM (ROSA NETO, 2002), entretien neuro psychologique et deux questionnaires élaborés pour la recherche. Résultats: sur l'échelle MMG on obtient un bon diagnostic tant sur le pré-test (84%) que sur le post-test (92%). Sur l'EDM, le domaine qui a obtenu la meilleure performance dans le pré-test a été l'organisation temporelle et dans le post-test, ce fût la motricité fine, cependant la plus mauvaise performance dans le pré-test a été l'organisation spatiale et dans le post-test le schéma corporel. Par l'application des questionnaires, on a observé un haut niveau de motivation et de satisfaction du patient. Conclusion: les résultats montrent que l'utilisation de la Nintendo WII sur l'amélioration de la DMD, a favorisé une augmentation des scores du développement des habilités motrices (organisation temporelle et motricité fine), en plus de favoriser la motivation du patient.

MOTS CLÉS: Nintendo WII, Dystrophie Musculaire de Duchenne, réhabilitatio

UTILIZACIÓN DE LA NINTENDO WII EN REHABILITACIÓN DE LA DISTROFIA MUSCULAR DE DUCHENNE.

RESUMEN

Introducción: La distrofia muscular de Duchenne (DMD) es un trastorno recesivo ligado al cromosoma X, y la deficiencia de un gen (Xp21) resulta en una producción alterada de la proteína distrofina . Evoluciona con la debilidad muscular progresiva. Objectivo : Determinar la eficacia del uso de la Nintendo Wii en la rehabilitación de la distrofia muscular de Duchenne . Metodo: Este estudio incluyó a paciente con DMD, varón, 10 años. La rehabilitación se utilizó Nintendo Wii Video Game. La evaluación se realizó a través de GMFM (Gross Motor Función Medida), EDM (Escala de Desarrollo Motor), entrevista neuropsicológica y dos cuestionarios diseñados para la investigación. Resultados: La escala GMFM, se obtuvo buen pronóstico, tanto en el pre-test (84%) como pos-test (92%). En EDM, el área con el mejor desempeño en la prueba preliminar era la organización temporal y posterior a la prueba, la motricidad fina y la organización temporal, mientras que el peor desempeño en la prueba preliminar era la organización espacial y esquema corporal post-test. A través de los cuestionarios , hubo un alto nivel de motivación y satisfacción del paciente. Conclusión: Los resultados mostraron que el uso de la Nintendo Wii en la rehabilitación de la DMD , lo que las puntuaciones más altas de desarrollo de las habilidades motoras (organización temporal y la motricidad fina) , además de mejorar la motivación del paciente .

PALABRAS CLAVE: Nintendo Wii , distrofia muscular de Duchenne, Rehabilitación .

UTILIZAÇÃO DO NINTENDO WII NA REABILITAÇÃO DA DISTROFIA MUSCULAR DE DUCHENNE.

RESUMO

Introdução: a Distrofia Muscular de Duchenne (DMD) é uma doença recessiva relacionada ao cromossomo X, sendo que a deficiência de um gene (Xp21) resulta em falha na produção da proteína distrofina. Evolui de forma progressiva com fraqueza muscular. Objetivos: verificar a eficácia da utilização do Nintendo Wii na reabilitação da Distrofia Muscular de Duchenne. Métodos: participou do estudo um paciente com DMD, sexo masculino, 10 anos. Para reabilitação foi utilizado o Vídeo Game Nintendo Wii. A avaliação foi realizada através do GMFM (Gross Motor Function Measure), EDM (Escala de Desenvolvimento Motor), entrevista neuropsicológica e dois questionários elaborados para pesquisa. Resultados: na escala GMFM, se obteve bom prognóstico, tanto no pré (84%) como pós-teste (92%). Na EDM, a área com melhor desempenho no pré-teste foi organização temporal e no pós-teste, motricidade fina e organização temporal; já o pior desempenho no pré-teste foi organização espacial e no pós-teste, esquema corporal. Através da aplicação dos questionários, observou-se alto nível de motivação e satisfação do paciente. Conclusão: os resultados demonstraram que a utilização do Nintendo Wii na reabilitação da DMD, proporcionou aumento nos escores de desenvolvimento das habilidades motoras (organização temporal e motricidade fina), além de favorecer a motivação do paciente.

PALAVRAS-CHAVE: Nintendo Wii, Distrofia Muscular de Duchenne, Reabilitação