

REVISTA NEUROCIÊNCIAS

ISSN 0104-3579
ISSNe 1984-4905

VOLUME 23 (Suplemento) 2015

Anais do I Simpósio Brasileiro de Investiga- ção de Doenças Neuromusculares

ESCOLA PAULISTA DE MEDICINA –
UNIVERSIDADE FEDERAL DE SÃO PAULO
(EPM/UNIFESP)

São Paulo, SP
25 e 26 de abril de 2015

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Anais do I Simpósio Brasileiro de Investigação de Doenças Neuromusculares

Data: 25 e 26 de abril de 2015

Local: Escola Paulista de Medicina – Universidade Federal de São Paulo (EPM/UNIFESP)

Organizadores: Dra. Francis Meire Fávero
Dr. Acary Souza Bulle Oliveira
Dra. Mariana Callil Voos
Dra. Cristina dos Santos Cardoso de Sá
Dra. Fátima Aparecida Caromano

Objetivo: I Simpósio Brasileiro de Investigação de Doenças Neuromusculares, destinado para reunir todos os pesquisadores, com o objetivo de trocar informações sobre vários aspectos das doenças neuromusculares, principalmente quanto as formas de avaliação e qualidades de vida.

PROGRAMA

25/04/2015 sábado (Anfiteatro Clóvis Salgado)

8h00 Entrega do material

Secretária

8h10 Abertura do evento

Prof. Dr. Acary Souza Bulle Oliveira

Prof. Dr. Fernando Morgadinho Santos Coelho

Prof. Dr. Ricardo M. Arida

8h30 Um breve histórico sobre a importância das Escalas no Brasil

Prof. Dra. Cristina dos Santos Cardoso de Sá - UNIFESP

8h50 Function Testing Manual Jebsen Taylor

Prof. Dra. Cristina dos Santos Cardoso de Sá - UNIFESP

9h30 Segmental Assessment of Trunk Control (SATCo)

Prof. Dra. Cristina dos Santos Cardoso de Sá - UNIFESP

10h10 Coffee break

10h30 Medidas da Função Motora - MFM

Prof. Dra. Cristina Iwabe - UNICAMP

11h10 Effects of a program based on home-exercises on muscle force and functional independence in patients with amyotrophic lateral sclerosis: a 2-year follow-up study

Prof. Dra. Maria Elisa Pimentel Piemonte - USP

11h50 Functional evaluation scale for Duchenne muscular dystrophy (FES-DMD)

Prof. Dra Michele Emy Hukuda - USP

12h30 almoço

13h30 Visual-cognitive-motor skills in children with spinal muscle atrophy - type I

Prof. Dra. Mariana Callil Voos - USP

14h10 Tarefas da realidade virtual para avaliação do tempo de Movimento em doenças Neuromusculares

Prof. Dr. Carlos Bandeira Monteiro de Mello - USP

14h50 Translation, cross-cultural adaptation and reliability of S.W.I.M. scale

Prof. Ms. Ana Angélica R. de Lima - USP

15h30 Coffee break

15h50 Escores e Escalas Diagnosticas em Ensaio Clínicos

Prof. Dr. Isac de Castro – USP

Apresentação Oral dos Pôsteres

26/04/2015 domingo (Anfiteatro Flávio da Fonseca)

8h30 Correlation between the 10 meters walking time and motor function measure in the Duchenne Muscular Dystrophy

Ivoneide PO Tozzo, Francis M Favero, Mariana C Voos, Fátima A Caromano, Cristina Iwabe, Acary SB Oliveira

9h10 Relationship between motor and cognitive change in patients with Duchenne Muscular Dystrophy

Kamila TF Dias, Mariana C Voos, Maria Elisa P Piemonte, Francis M Favero, Fátima A Caromano, Jecilene R Costa-Frutoso, Acary SB Oliveira

9h50 Functional performance in reality of virtual training in patients with Amyotrophic Lateral Sclerosis

Ana Carolina C Santos, Francis M Favero, Maria Elisa P Piemonte, Carlos BM Monteiro, Isabela L Trevisa, Talita D Silva, Acary SB Oliveira

10h10 Coffee break

10h30 Application of the portuguese version of the Jebsen Taylor test for patients with limb-girdle muscular dystrophy

Mariana A Rays, Mariana C Artilheiro, Francis M Favero, Fátima A Caromano, Acary SB Oliveira, Cristina SC Sá

11h10 Correlation between Medical Research Council (MRC) and the timed walking in the patients with Duchenne Muscular Dystrophy (DMD)

Alessandra C Santana, Francis M Favero, Mariana C Voos, Cristina SC Sá, Fátima A Caromano, Acary SB Oliveira

11h50 Aprendizagem motora por meio de jogo de labirinto em telefone móvel na Paralisia Cerebral

Juliana N Paula, Camila M Capelini, Talita D Silva, Francis M Favero, Vitor Kagohara, Luiz C Abreu, Carlos BM Monteiro

12h30 almoço

13h30 Characterization of muscle force on Brazilian sample of patients with limb-girdle muscular dystrophy type 2 (calpainopathy)

Jéssica G Marin, Mariana C Voos, Isabela PA Leite, Francis M Favero, Acary SB Oliveira, Fátima A Caromano

14h10 Clinical characteristics in the autosomal recessive limb-girdle Muscular Dystrophy - Sarcoglycanopathy: a report of cases

Geisa SC Moraes, Isabela PA Leite, Francis M Favero, Michele E Hukuda, Fátima A Caromano, Acary SB Oliveira, Mariana C Voos

14h50 Literature review on the assesment tools that showed Responsiveness in Neuromuscular Diseases

Tatiana AS Veríssimo, Michele E Hukuda, Francis M Favero, Acary SB Oliveira, Mariana C Voos, Fátima A Caromano

15h30 Coffee break

15h50 Utilização da realidade virtual para avaliar o desempenho do ombro de pacientes com Lesões do Manguito Rotador

Alexandre F Alves, Francis M Favero, Abrahão AJ Quadros, Talita Dias, Carlos BM Monteiro, Acary SB Oliveira, Isabela L Trevizan

16h30 Brief Report e Discussão

20:00 Encerramento

I Simpósio Brasileiro de Investigação de Doenças Neuromusculares

Na prática clínica, as doenças neuromusculares são de difícil diagnóstico, devido à falta de informações da evolução clínica ou métodos diagnósticos acessíveis e conclusivos. Na atualidade, os métodos adotados para a realização do diagnóstico são relacionados com a descrição dos sinais e sintomas, a evolução da doença, e a topografia; em poucos casos existem exames imuno-histopatológicos e moleculares que permitem diagnóstico mais conclusivo.

O presente Simpósio objetivou apresentar os mais recentes trabalhos na área de Doenças Neuromusculares. Na busca incansável de um diagnóstico precoce correlacionando o melhor entendimento e a compreensão das características destas doenças, juntamente com os aspectos clínicos e exames complementares, sobretudo com avanços científicos mais recentes, os pesquisadores têm experimentado mudanças significativas nas abordagens de avaliação, com diagnósticos cada vez mais precisos e precoces, e abordagens terapêuticas mais apropriadas.

Para o seguimento e análise de respostas terapêuticas são necessários instrumentos de avaliação adequados, destacando-se as escalas funcionais, que devem ser sensíveis tanto para diagnóstico, quanto para prognóstico, e principalmente, para direcionamento de terapias multidisciplinares.

Dra. Francis Meire Fávero

Dr. Acary Souza Bulle Oliveira

Dra. Mariana Callil Voos

Dra. Cristina dos Santos Cardoso de Sá

Dra. Fátima Aparecida Caromano

**Organizadores do I Simpósio Brasileiro de Investigação
de Doenças Neuromusculares**

Function Testing Manual Jebsen Taylor

Cristina dos Santos Cardoso de Sá

Fisioterapeuta, Profa. Dra. Departamento Ciências do Movimento Humano, Curso de Fisioterapia, Universidade Federal de São Paulo.

The muscular dystrophies (MD) are a group of genetically determined disorders heterogeneous, identified clinically by great variety of phenotypes and severity and characterized by muscle weakness¹. Most MD share the same pathological feature. As the disease progresses, muscle tissue suffers injuries that may cause fibrosis formation and subsequently the muscle tissue is replaced by fat. However, the mechanism of evolution, and the speed at which it occurs in each of the specific forms may vary¹.

The progressive degeneration of skeletal muscle causes the muscle weakness and atrophy and contractures and deformities installation of the axial skeleton muscles, shoulder girdle and upper limb (UL) are inherent characteristics of all the MD group. It is known that the degree of the involvement of these musculature and injury may vary^{2,3}, however, the specific involvement of the MS in each of the described forms of the disease is poorly investigated.

The evaluation of upper limb function in MD is a useful clinical tool of great importance to the knowledge of functional limitations and to characterize the natural history of this disease group^{3,4}. The determination of appropriate measurements that reflect improvement or stabilization in functional ability is a challenge for professionals who follow patients with these types of neuromuscular disease (MND)^{4,5}.

The validated instruments applied in the MD population have been used in the assessment of motor function of the upper limbs, such as questionnaires Activity Limitations in Patients with Neuromuscular Disorders (ACTIVILIM) and ABILHAND, and scales Muscular Dystrophy Functional Rating Scale, Brooke and Measurement Motor function (MFM).

The Jebsen-Taylor test (TJT) developed in 1969 with the objective of evaluating the manual function commonly used in daily life activities and measure objectively the performance of functional tasks. Not only evaluates the distal aspect because there are too proximal region of action requirement of UL for the execution of tasks. It consists of seven tests: writing, turn card (simulate the act of turning the page), pick up small common objects, simulate power, take maid of game pieces, pick up large objects and light and pick up large objects and weighing⁶. The TJT features as a way to score the time

obtained by the volunteer to run the subtests. Continuously documents the manual skill, it's easy and quick to administer and use readily available materials⁷. The test was applied previously in 23 boys, wheelchairs and walkers, with DMD⁸.

The main objective of the study was to verify the applicability of TJT the MD in order to measure the manual function of individuals with motor impairments resulting from the MD and the ability to monitor the progression of this commitment over the course of the disease.

The results revealed that the TJT measures the manual function of MD, and also indicates the need to include subtests assessing the activity mouth hand and bimanual activity for this population.

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Segmental Assessment of Trunk Control (SATCo)

Cristina dos Santos Cardoso de Sá

Fisioterapeuta, Profa. Dra. Departamento Ciências do Movimento Humano, Curso de Fisioterapia Universidade Federal de São Paulo.

Neuromuscular diseases are genetic diseases hereditary and progressive and represent a range of disorders that compromise the motor unit, the cell body of the neuron lower, its extension, the neuromuscular junction or skeletal muscle tissue¹. Among these diseases, muscular dystrophies (MD) are characterized by progressive and irreversible present with muscle tissue degeneration caused by the loss of several muscle proteins². Among the MD, there is great variety in the clinical severity of symptoms: some cases have early onset and rapid progression; However, there are cases in which the onset is slow and development is more slower³.

Muscle imbalances trunk present in individuals affected by DM, triggered by continued weakness, account for abnormal posture caused by imbalances in the paravertebral and abdominal musculature, which are responsible for providing stability to the body and greater postural control⁴.

Postural control is responsible for the action of gravity resistance and the body balance maintenance; It involves adjustments in order to maintain the aligned segments during body movement and according to the desired task⁵.

It is known that postural control of the trunk is essential for the implementation of various functional activities. Some studies have shown that the delay or defective development of postural system may restrict the ability of individuals to develop independence in mobility and manipulative activities^{6,7}. In addition, the loss in stem generates control compensation and deformities in the spine, which is very frequent in patients with muscular dystrophies. These changes in physiological spine curvatures hinder the achievement of thicker manuals and functions skills such as sitting and walking⁸.

The use of trunk control assessment scales in diabetic patients, allows the physiotherapist to draw up an appropriate treatment plan is directed to the specific area of deficit, in order to slow down the evolution of the loss of motor function in these patients

The Segmental assessment of trunk control (SATCo)⁹ is a validated test, internationally recognized and has been shown to be an excellent method of assessment¹⁰, as it evaluates the various segments of the trunk needed so that there is coordination and trunk control in

sitting posture¹¹. This scale allows to evaluate from the individual who has just head control to one who has total control of the trunk in relation to the static active and reactive control.

This study aimed to verify the trunk control level in patients with various muscular dystrophies, in varying degrees of disease progression, defined Vignos the range and types of column deformity of such patients.

The results showed that the patients with different degrees of disease progression (Vignos) had the same degree of control of trunk measured by SATCo. The scale allows SATCo separately evaluate the various segments of the trunk needed so that there is coordination and control of the trunk in a sitting posture, and the use of this scale can complement the assessment methods used in the rehabilitation of patients with muscular dystrophies.

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Motor Function Measure (MFM) scale applicability in Neuromuscular Diseases

Cristina Iwabe

Pós Doutora em neurologia FAPESP/UNICAMP, docente do curso de fisioterapia da Faculdade Integrada Metropolitana de Campinas, pesquisadora voluntária do grupo de estudos em doenças neuromusculares UNICAMP.

Neuromuscular Diseases (NMD) include a group of disorders which primary disease affects any part of the motor unit from the anterior horn cells of the spinal cord to the skeletal muscle. Among the different clinical pictures presented, these diseases occur with muscle weakness topography and varying severity. The elucidation of the severity of functional impairment is crucial to the prognosis and targeting of therapeutic approaches, as the motor execution capacity can vary even in individuals of the same family with the same disease in different age groups (Rocco et al., 2005; Iwabe, Pfeilsticker and Nucci, 2009). It is important to consider that in patients with NMD functional loss may be related to the presence of muscular retractions and / or due to a relatively stable pattern of muscle strength, as grade 4 on the scale Medical Research Council (Linssen et al., 1997). Faced with the need for accurate correlations between muscle weakness and motor function, the use of objective parameters such as specific and validated functional scales for analysis of evolution, topography and prognosis of patients with NMD is critical for targeting the therapeutic treatment. Among the instruments used can cite Vignos and Brooke scale, Jebsen hand function test, Hammersmith and Egen Klassification, but these are validated for a particular type of NMD, mainly Duchenne muscular dystrophy, it is

not possible to evaluate other neuromuscular disorders as hereditary neuropathies. To this purpose, in 1998 was created the scale Motor Function Measure (MFM) by the French group MFM, led by Dr. Carole Bérard, which aims to outline the symptoms and evolution of all NMD and direct goals of therapeutic measures and rehabilitation encompassing patients, aged 2 to 6 years of age (MFM-20 scale) to 60 years (MFM-32). MFM includes 32 items (for patients over 6 years to 60 years of age) or 20 items (children aged 2 to 6 years of age) which evaluate the overall functionality of individuals, divided in three dimensions: D1 (standing position and transfers), D2 (axial and proximal motor function) and D3 (distal motor function), where each item is graded on a 4-point scale (0-3). In 2011, the scale has been translated and validated in Brazil, the group of NMD at Unicamp, and the physiotherapist Prof. Dr. Cristina Iwabe being the reference to trainees and capacity other professional in this country. Currently the MFM has been used in several scientific studies to identify the severity of a given NMD, tracking the progress of each patient and cohort design of genotype and phenotype correlations, the correlation between other diagnostic methods, to delineate the prognosis as the loss of functional capabilities and evaluate the efficacy of therapeutic approaches.

Effects of a program based on home-exercises on muscle force and functional independence in patients with amyotrophic lateral sclerosis: a 2-year follow-up study

Maria Elisa Pimentel Piemonte

Department of Physiotherapy, Speed and Occupational Therapy of the Medical School - University of São Paulo

Currently, there is a considerable number of evidences from animals (Carreras et al. 2010) and human studies (Lui AJ & Byl NN, 2009) showing the positive effects of moderate intensity exercises on Amyotrophic Lateral Sclerosis (ALS). These evidences emphasize the important role of Physical Therapy as part of ALS treatment (Majmudar S; Wu J; Paganoni S, 2014). However, the physical incapacity, associated to social/ economic problems prevent the access to physical therapy services. In face this, we developed a study that aimed to investigate the effects of a program based on home exercises on muscle force and functional independence in ALS patients. Twenty eight patients (mean age 51.4 years; 3273 years) were divided in two groups, according to their availability, to follow the programs: Experimental Group (EG) and Control Group (CG). Inclusion criteria included the absence of severe depression (Beck Depression Inventory) and of cognitive impairment (Mini mental State Examination). Patients were assessed at the beginning of the study and 6, 12, 18 and 24 months after with ALS Functional Rating Scale (ALSFRS), for functional inde-

pendence and Kendall Test (KT), for muscle force. EG received detailed explanation about each exercise and was asked to perform a sequence of exercises at home once a day. Exercises were developed to prevent the progressive functional loss and were selected according to the stage of ALS of each patient. The performance in the exercises was reviewed monthly. CG received only general information about the disease and the risks of sedentarism. The results analyzed by 2 (group) X 5 (assessments) repeated measures ANOVAs, showed a statically significant interaction between the factors for ALSFRS scores [p .90]. Post hoc comparisons using the Tukey HSD test indicated that there was significant loss of functional independence for CG only. In contrast, there was only a significant effect of assessments for KT scores [p .90]. Post hoc comparisons using the Tukey HSD test indicated that there was significant loss of muscle force independently of group. The home exercises was able to prevent the loss of functional independence for 2 years in patients with ALS, regardless of the progressive loss of muscle force.

Functional evaluation scale for Duchenne muscular dystrophy (FES-DMD)

Michele Emy Hukuda

PhD, University of São Paulo, School of Medicine, Laboratory of Physiotherapy and Behavior

Motor assessment includes the analysis of muscle strength and joint mobility. However, this evaluation is not enough for understanding the functional limitations because, as DMD progresses, the child employs compensatory movements and postural adaptations to keep performing daily life activities. The evaluation of dynamic functional activities such as walking, climbing up and down stairs, sitting and rising from a chair or the ground, can be performed by direct or indirect observation (films). Therefore, standardized protocols are necessary for observing and recording information about the movements involved on each activity. However, more sophisticated methods of movement assessment, such as electromyographic and kinematic analyses are accurate, but expensive and difficult to be performed in the clinical practice. A detailed description of the daily life compensatory movements, focusing on the remaining strategies available, was the basis for the development of the Functional Evaluation Scale for patients with DMD (FES-DMD). This scale allows qualitative and descriptive analyses of the patterns of compensatory movements during functional tasks and the detection of important changes in muscular synergies. It is composed of four do-

main: domain 1 (D1) - sitting and rising from a chair; domain 2 (D2) - gait; domain 3 (D3) - going up and down stairs and domain 4 (D4) - sitting and standing from the ground. FES-DMD has been designed to be used on a clinical routine evaluation, in which the patient is filmed performing functional activities. The film enables the observation of the movements on the frontal and sagittal planes. This routine decreases the assessment time and creates an image bank. The score is reliable and most ICC and kappa values varied from very good to excellent, showing reliability. The assessment with FES-DMD involves watching the films and, simultaneously, filling charts and recording data. It is also possible to generate partial and total scores of the activities, measure and record the time spent during each activity. To facilitate these tasks we developed the software that displays on the computer screen, simultaneously, the film for observation and the chart to fill. The software, which has been called FES-DMD-DATA, uses Brazilian Portuguese. Its usability has been demonstrated. It refers to the speed with users can learn to use some tool, their efficiency in using it, their degree of error and how much they enjoy using it.

Visual-cognitive-motor skills in children with spinal muscle atrophy - type I

Mariana Callil Voos

University of São Paulo, Medical School, Physiotherapy Department (São Paulo - SP, Brazil). E-mail address: ftmarivoos@gmail.com

Due to their severe motor paralysis and poor oral communication, it is difficult to evaluate and follow children with spinal muscle atrophy type I (SMA-I, Werdnig-Hoffman). Studies have shown contradictory findings about cognitive skills in SMA-I. Genetic alterations and the lack of environmental interaction due to the severe paralysis may interfere with cognitive development. We aimed to (1) propose the use of TOBII-PCEye, (2) investigate the visual-cognitive-motor performance of children with SMA-I, (3) compare children with SMA-I to healthy controls. Twelve children (3-10 yrs old) with SMA-I and 12 matched controls were assessed. Children with SMA-I were positioned on adapted wheelchair, facing the notebook with TOBII-PCEye. Figures and colors association tasks were proposed for all children. For the 7-10 yrs old children numbers and letters association tasks were also proposed. The figures were displayed in 4x4cm squares. The response required was selecting each correct pair of

squares by 2000ms eye fixation. The number and percentage of correct answers was registered. ANOVAs compared the scores of the groups. Children with SMA-I were also assessed with Chop Intend (CI), Pediatric Evaluation of Disability Inventory (PEDI) and Pediatric Quality of Life Inventory (PedsQL). TOBII-PCEye quantified the visual-cognitive-motor performance of the children with SMA-I. Correct answers varied from 0% to 100% with a median of 40% correct answers on the visual-cognitive-motor task for children with SMA-I and 70% for controls ($p<0.05$). Children with SMA-I scored 3-40% on CI, 5-20% on PEDI and 29-59% on PedsQL. Despite having low motor performance, functional independence and quality of life, children aged 4 yrs or older could perform TOBII-PCEye tasks. The figures association difficulty, shown by many children with SMA-I, suggests that the genetic alterations and the restricted environmental interaction can contribute to cognitive impairment.

Translation, cross-cultural adaptation, and reliability of S.W.I.M. scale

Ana Angélica Ribeiro de Lima

Mestranda pelo Programa Ciências da Reabilitação da FMUSP, Especialista na área de Fisioterapia Aquática.

Assessment tools are fundamental for basis for scientific research and determine clinical objectives with logical reasoning, based and appropriate to the population to be treated. In aquatic therapy, these instruments in Portuguese are rare. The translation and cross-cultural adaptation process of scale allows an increase in number of searches and greater equivalence between original scale and scale to be translated.

According to translation and cross-cultural adaptation guides for scales, it is suggested translation, translation synthesis, back-translation, the committee synthesis of expert, finally the pre-test phase.

Subsequently, carrying out intrate and intra-reliability method, it is another feature to attribute quality of scale that has been translated.

The Swimming with Independent Measure (SWIM) used in aquatic therapy area, is a scale to measure the water skills of children with physical disabilities or learning and direct intervention programs in the aquatic environment. The original scale in English, was translated, cross-culturally adapted for Brazilian Portuguese and evaluated by reliability method, with equivalence to the original version.

Apresentação Oral de Posters

Correlation between the 10 meters walking time and motor function measure in the Duchenne Muscular Dystrophy

Ivoneide Paula de Oliveira Tozzo¹, Francis Meire Favero¹, Mariana Callil Voos², Fátima Aparecida Caromano², Cristina Iwabe³, Acary Souza Bulle Oliveira¹

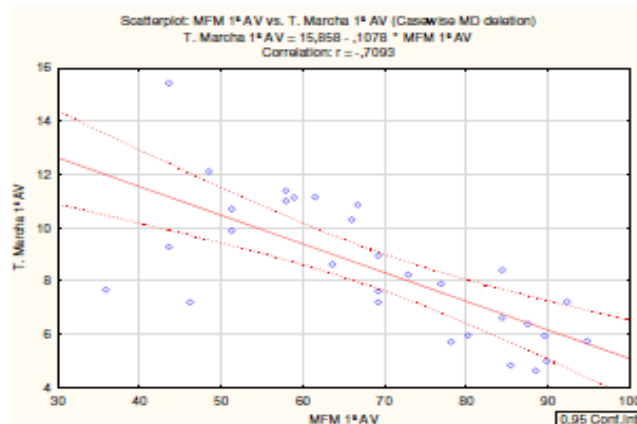
1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

3.Universidade de Campinas (UNICAMP), Campinas-SP, Brasil.

Introduction. The Duchenne Muscular Dystrophy (DMD) is a neurodegenerative disease that affects mainly motor functions. Nowadays it is necessary to study tools to evaluate and classify, as well as the scale of motor function measure (MFM) that is widely used to measure the motor and functions capabilities, among other measures as the timer that can assist in better understanding of the evolution this disease. **Objective.** To correlate the time (seconds) measured in a 10 meters walking and the motor function of patients with DMD. **Method.** The retrospective study covers the period 2010 to 2013. Data was collected directly from the records of evaluations of patients with diagnosis of DMD, the research Division of Clinical Investigation on Neuromuscular Diseases (DCINM), Department of Neurology and Neurosurgery of Federal University of São Paulo. In all evaluated patients the MFM scale was used as a tool to measure the 10 meters walk test in a normal speed. This study may contribute to the use of an accurate measurement and rapid implementation to check the progression of the disease. Results: The records of 30 patients with DMD were analyzed and relevant information was used, such as: age, defined diagnosis, Vignos to check the staging disease, time in seconds of 10 meters walk and the MFM. Pearson correlation analysis was performed in the results of the total percentage of MFM and the 10 meters walk time. In this study we observed that there is a strong relationship between the values, $r=0.709$ ($p<0.05$). **Conclusion.** Our data suggests that there is a strong correlation between the time of the 10 meters walk, in seconds, and the MFM scale, we can conclude that both measures can be used to evaluate the disease progression. It was observed that the measured time is a reliable tool to track the motor function evolution of patients with DMD.

Figure 1. The analysis was done with Pearson correlation test, investigating the relationship between the measured time (seconds) and the walk on 10 meters with the MFM. Being AV: evaluation. The equation that best represents the straight set the point cloud is $\text{time}=15.86-.11\text{XMF}$. Correlation coefficient: $r=0.71$ ($p<0.05$). The confidence interval is represented by the dotted INES and calculated from the squared correlation coefficient (r^2).



Keywords. Duchenne Muscular Dystrophy, Muscle Strength, Measures, Motor Activity, Function.

Relationship between motor and cognitive change in patients with Duchenne Muscular Dystrophy

Kamila Teixeira Foiani Dias², Mariana Callil Voos¹, Maria Elisa Pimentel Piemonte¹, Francis Meire Favero², Fátima Aparecida Caromano¹, Jecilene Rosana Costa-Frutoso³, Acary Souza Bulle Oliveira²

1.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

2.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

3.Coordenadora do Programa de Pós-Graduação em Ciências da Saúde - Universidade Ibirapuera (UNIB), São Paulo-SP, Brasil.

Introduction. Duchenne Muscular Dystrophy (DMD) have motor and respiratory involvement also may have cognitive impairments such as difficulties in visuospatial abilities, language, working memory and phonological therapy processes. **Objective.** To describe the motor and cognitive changes in a group of patients with DMD and to investigate possible relationships between cognitive and motor performance. **Method.** Cross-sectional, observational study in Division of Clinical Investigation on Neuromuscular Diseases (DCINM), Department of Neurology and Neurosurgery of Federal University of São Paulo. Tests were performed evaluating cognitive and motor function of patients who were 10 to 30 years. Each patient underwent a protocol consists of five tests: 1.

Mini Mental State Examination (MMSE), 2. Watch Drawing, 3. verbal fluency, 4. digit Order, 5. Battery Brief Cognitive Screening. Patients also underwent a driving assessment with application of motor function measure (MFM) and Vignos scales. **Results.** The Spearman correlation tests to investigate possible relationships between cognitive and motor variables. Was adopted alpha <0.05 significance level. There were correlations ($r>0.70$ and $p<0.05$) between MFM and Vignos ($r=0.75$, $p<0.05$) between MMSE and education ($r=0.70$, $p<0.05$); between MMSE and clock test ($r=0.83$, $p<0.05$); with MMSE and digits - direct order ($r=0.76$, $p<0.05$) and digits - reverse order ($r=0.75$, $p<0.05$). Also showed correlations between clock test and digits - reverse order ($r=0.75$, $p<0.05$); between digits - direct order and digits - reverse order ($r=0.76$, $p<0.05$). Following the strong correlations ($r>0.70$ and $p<0.05$) are also presented through scatterplots. **Conclusion.** There was variability in cognitive and motor performance of patients with DMD. Correlations were observed between cognitive variables (education vs. MMSE, MMSE vs. Clock Test; MMSE vs. digit direct order; MMSE vs. digit reverse order, clock test vs. digit reverse order, digits direct order vs. order reverse). There was a correlation between motor variables (MFM vs. Vignos). However, there were no correlations between cognitive and motor variables.

	Age	School	MMSE	CT	DD	DR	Animals	P	MFM	Vignos
Average	14.1	6.0	21.5	5.7	5.5	2.4	10.6	5.0	0.54%	5.0
Min	5	1	2	0	0	0	0	0	0.15%	1
Max	27	12	30	10	10	7	18	14	0.90%	9
DP	4.2	3.8	7.6	3.8	2.7	2.0	4.5	3.7	0.21%	2.6

Table 1. Description of the sample characteristics and cognitive and motor variables. Medium, minimum values (Min), maximum (Max) and standard deviation (SD) of age, education, Motor Function Measure (MFM), Vignos Scale, Mini-Mental State Examination (MMSE), clock test (CT), digits direct order (DD), digits reverse order (DR), words with the letter p (P), enumeration of animals.

	Age	School	MFM	Vignos	MMSE	CT	DR	DD	P	Animals
Age	1.00	0.46	-0.33	0.52	0.44	0.31	0.22	0.30	0.21	0.59
School	0.46	1.00	0.19	0.07	0.70	0.67	0.56	0.54	0.59	0.53
MFM	-0.33	0.19	1.00	-0.75	0.21	0.32	0.19	0.21	0.21	0.11
Vignos	0.52	0.07	-0.75	1.00	0.08	0.01	0.20	0.18	0.09	0.27
MMSE	0.44	0.70	0.21	0.08	1.00	0.83	0.76	0.77	0.70	0.66
CT	0.31	0.67	0.32	0.01	0.83	1.00	0.76	0.68	0.66	0.59
DR	0.22	0.56	0.19	0.20	0.76	0.76	1.00	0.76	0.58	0.47
DD	0.30	0.54	0.21	0.18	0.77	0.68	0.76	1.00	0.64	0.56
P	0.21	0.59	0.21	0.09	0.70	0.66	0.58	0.64	1.00	0.64
Animals	0.59	0.53	0.11	0.27	0.66	0.59	0.47	0.56	0.64	1.00

Table 2. Correlation between cognitive and motor variables (r values). For all correlations with $r>0.40$, there was obtained $p<0.05$. Motor Function Measure (MFM), Mini Mental State Examination (MMSE), clock test (CT), digits direct order (DDO), digits reverse order (DRO), words with the letter p (P), enumeration of animals (animals).

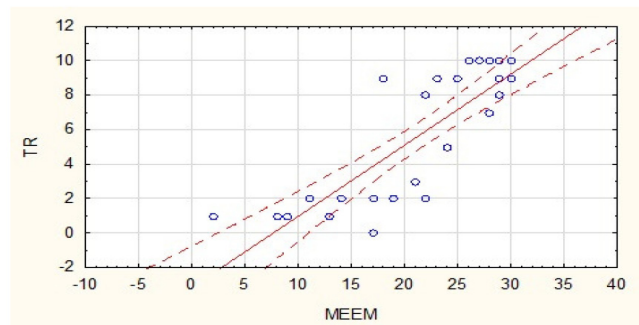


Figure 1. Correlation between performance on the clock test (CT) and the Mini Mental State Examination (MMSE) ($r=0.83$, $p<0.05$).

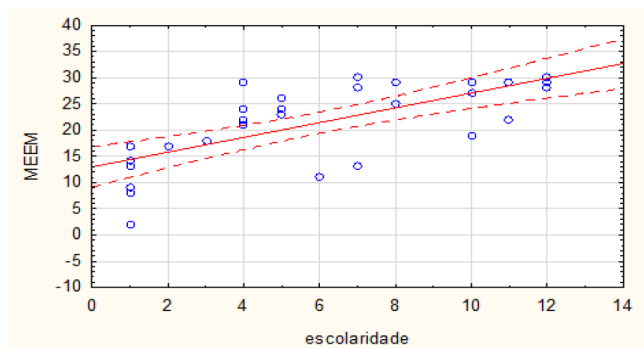


Figure 2. Correlation between performance on the Mini Mental State Examination (MMSE) and education (number of years of formal study) ($r=0.70$, $p<0.05$).

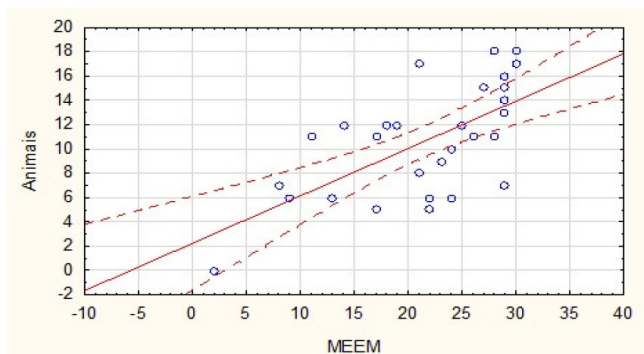


Figure 3. Correlation between performance on the Mini Mental State Examination (MMSE) and the verbal fluency test - animals ($r=0.65$, $p<0.05$).

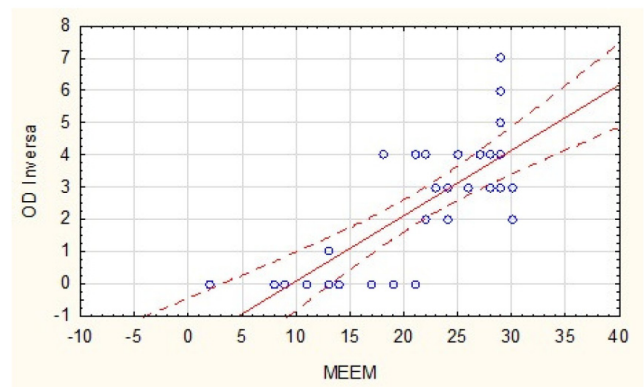


Figure 4. Correlation between performance on the Mini Mental State Examination (MMSE) and the digit test - reverse order ($r=0.75$, $p<0.05$).

Functional Performance In Reality Of Virtual Training In Patients With Amyotrophic Lateral Sclerosis

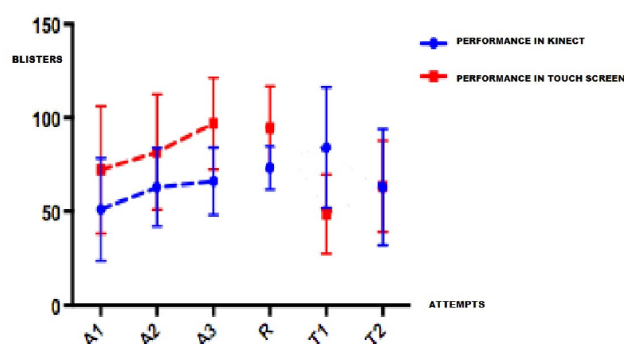
Ana Carolina Costa Santos¹, Francis Meire Favero¹, Maria Elisa Pimentel Piemonte², Carlos Bandeira de Mello Monteiro², Isabela Lopes Trevisa², Talita Dias da Silva¹, Acary Souza Bulle Oliveira¹

1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

Introduction. The Amyotrophic Lateral Sclerosis (ALS) becomes a challenge for researchers looking for tools which might assist in the development of new methods of diagnosis and evaluation, however, technologies such as the usage of games in virtual environments might also be used as a resource for review functional. **Objective.** Analyze the performance of the functionality and virtual training in patients with ALS. **Method.** Was selected individuals in Division of Clinical Investigation on Neuromuscular Diseases (DCINM), Department of Neurology and Neurosurgery of Federal University of São Paulo. Inclusion and exclusion criteria were applied, all patients in the study were evaluated by ALSFRS scales, FSS and AL-SAQ-40. The software for the games was created in partnership with the Travel Team Information Systems School of Arts, Sciences and Humanities, called "TUCANO GAMES". For the purposes of the game it was used 3 interfaces Kinect, the Touch Screen and the Leap Motion. Data analysis were performed with the dependent variables subjected to ANOVA with factor 2 (Groups: Kinect x Touch Screen) for 2 times of acquisition with repeated measures in the last 2 factors. Linear regression. **Results.** The study included 20 subjects with ALS. Increasing of broken blisters in the first attempt (A1) (m=62) for the last attempt (A3) (m=81) with better performance on the touch screen (m=85) than in kinect (m=58). There was no difference between the last attempt (A3) (m=81) and retention (m=84), Both groups showed greater amount of broken blisters (m=84) compared to the transfer (m=66), however this difference was more pronounced in the group which began on the touch screen (retention phase) (m=95) and was for Kinect (m=49), while for the group which began in Kinect, this difference did not occur (m=73 and 84, respectively). In transfer to Leap motion, there was no difference between attempts which both groups performed better on retention (m=84) than in the transfer (m=63) Linear regression showed a significant regression model, resulting in the following equation: improvement. **Conclusion.** It was observed

Figure 1. A1-A3: attempts of being acquired; R=retention trial phase; T=transfer attempt opposite phase to the interface used in the acquisition and retention stage; T2=the transfer phase attempt to Leap Motion.



better performance on the touch screen at all stages of the game. As for functionality, it was concluded that this type of software might be used as an assessment of disease progression compared the scores made in the game and the score in the functional scales.

Keywords. Amotrophyc Lateral Sclerosis, Motor Neuron Disease, Measures, Motor Activity, Function

Application of the portuguese version of the Jebsen Taylor test for patients with limb-girdle muscular dystrophy.

Mariana de Abreu Rays¹, Mariana Cunha Artilheiro², Francis Meire Favero¹, Fátima Aparecida Caromano², Acary Souza Bulle Oliveira¹, Cristina dos Santos Cardoso de Sá¹

1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

Introduction. The Limb-girdle muscular dystrophy type (LGMD) is a heterogeneous group with great variability of genetic disruptions, where muscle weakness is predominant in the pelvic and shoulder girdles. **Objective.** This study aimed to determine the responsiveness of the Jebsen Taylor scale (JTT) in patients with LGMD, identify the performance variability in each test and its compensations through the Functional Ability Scale (FAS). **Method.** The sample consisted of eight patients males diagnosed with LGMD that conducted tests as JTT and FAS, in addition to instruments used in the literature to evaluate motor and cognitive aspects such as Vignos and the Mini Mental State Examination (MMSE). **Results.**

Was found that the higher the score at the Vignos scale, the greater the time spent for the execution of the subtests of JTT, and a great variety of times probably occurred by heterogeneity sample. The FAS proved to be a good tool to assess disease progression and compensations made by the sample. The scale showed at the revaluations that the compensations related to flexion, extension, trunk rotation and inclination and shoulder elevation and abduction increased in frequency. In addition to describing new compensations to the distal region of the upper limb. **Conclusion.** The JTT along with the FAS allowed a quantitative and qualitative evaluation of the sample.

Keywords. Muscular Dystrophy, Muscular Dystrophy Limb-Girdle, Upper Extremity, Evaluation, Disability Evaluation

Correlation between Medical Research Council (MRC) and the timed walking in the patients with Duchenne Muscular Dystrophy (DMD)

Alessandra Corrales de Santana¹, Francis Meire Favero¹, Mariana Callil Voos², Cristina dos Santos Cardoso de Sá¹, Fátima Aparecida Caromano², Acary Souza Bulle Oliveira¹

1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP). São Paulo-SP, Brasil.

Introduction. Duchenne Muscular Dystrophy (DMD) is the most common and severe form of dystrophy, with the evolution of the disease the loss of muscle strength and motor function become evident, with subsequent loss of the walk. The walking is one of the main motor function, representing an important role in the clinical course of the disease, it is essential to indicate the progression of the disease. Currently, there are some evaluation tests using the time with form of accurate measurement, which is easily accessible and quick applicability, another muscle strength test or Medical Research Council (MRC), can consider these two main measures to analyze the functional loss. **Objective.** Compare the Medical Research Council (MRC) and the time in seconds in the walk of 10 meters in patients with DMD. **Method.** Analyzed 86 records in the Division of Clinical Investigation on Neuromuscular Diseases (DCINM), Department of Neurology and Neurosurgery of Federal University of São Paulo, collected diagnostic data, muscle strength or the Medical Research Council (MRC) of the lower limbs and the time in seconds of the walk in a distance of 10 meters. The analysis was performed using

the Pearson correlation test, investigating the relationship between the measured time (seconds) of the walking on 10 meters with the Medical Research Council (MRC). Significant were those whose value $p < 0.05$. **Results.** The study included the records of patients with DMD who had all the information necessary for performing the correlation ($n=30$). Being Division of Clinical Investigation on Neuromuscular Diseases (DCINM), Department of Neurology and Neurosurgery of Federal University of São Paulo, the average age was 10.86 ± 3.18 , all confirmed with DNA analysis. By analyzing the values obtained from the statistical analysis, we found correlations between the proximal muscle strength of the lower limbs (PMSLL) and the walking test of 10 meters ($r = -0.47$; $p < 0.05$) and the distal muscle strength lower limb (DMSLL) and gait test of 10 m ($r = -0.68$; $p < 0.05$). **Conclusion.** Regarding the results presented in this study, it is concluded that there is a correlation between the variables, measured time of travel at 10 meters and the muscle strength, especially in DMSLL. From these clinical findings, it is possible to infer the time that the patient may have to walking from the muscle strength and vice versa.

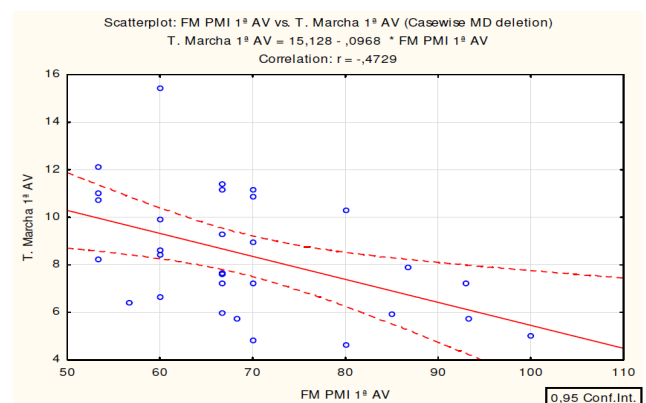


Figure 1. The analysis was done with Pearson correlation test, investigating the relationship between the measured time (seconds) of the march held on 10 meters and the proximal muscle strength MI. Being AV: evaluation. The equation that best represents the straight set the point cloud is $\text{time} = 15.13 - .97 \times \text{FMP}$.

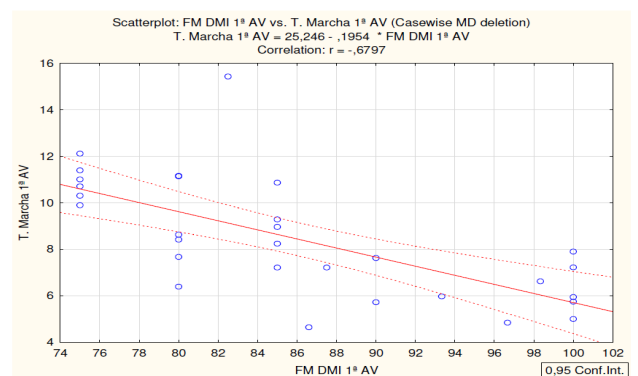


Figure 2. The analysis was done with Pearson correlation test, investigating the relationship between the measured time (seconds) of the march held on 10 meters and the distal muscle strength MI. Being AV: evaluation. The equation that best represents the straight set the point cloud is $\text{time} = 25.25 - 0.19 \times \text{FMD}$.

Motor Learning for maze game in Mobile Phone in Cerebral Palsy

Juliana Nobre de Paula¹, Camila Miliani Capelini², Talita Dias da Silva¹, Francis Meire Favero¹, Vitor Kagohara³, Luiz Carlos de Abreu², Carlos Bandeira de Mello Monteiro²

1.Universidade Federal de São Paulo-UNIFESP, São Paulo-SP, Brasil.

2.Faculdade de Medicina da Universidade de São Paulo - FMUSP, São Paulo-SP, Brasil.

3.Escola de Artes Ciências e Humanidades da Universidade de São Paulo-EACH, USP, São Paulo-SP, Brasil.

Introduction. Cerebral Palsy (CP) is a group of permanent disorder of posture and movement development, causing limitations in activities that are assigned to a non-progressive disorder that occurs in fetal brain development or infancy. A current movement option and using technological advances are tasks in mobile phone that can be an option for intervention in the recovery in various populations with sensory and motor abnormalities.

Objective. To verify the occurrence of motor learning in a maze game task on mobile phone in individuals with PC. **Method:** A total of 50 individuals, 25 with medical diagnostic PC (GE) and 25 subjects with typical development (TD), matched for age and sex. The execution of the task was to play a maze on a mobile phone. **Results.** The subjects performed 20 repetitions of the maze game in the acquisition phase and 5 repetitions in the phases of retention and transfer. A repeated measures ANOVA was used to compare the blocks (first-and last A1-A4 blocks the phase acquisition, retention-and transfer R-T) and Groups (Cerebral Palsy and Control). **Conclusions.** It has been found that during the acquisition phase in the EG was improved performance and consequently the reduction time, taking into consideration the average of the first and last attempt. However, this performance was lower than the control group, confirming the interference of the motor change the PC on performing motor skills. In the retentive phase there was no effect or interaction to note indicating the time in comparison to the acquisition phase was maintained despite the higher GE time reference to GC. Regarding the transfer both groups had long compared to the retention and GE continued to show long while performing the task.

Keywords. Motor Activity, Cerebral Palsy, exposure therapy to virtual reality, Mobile Phone, Motor Skills

Characterization of muscle force on Brazilian sample of patients with limb-girdle muscular dystrophy type 2 (calpainopathy)

Jéssica Gomes Marin¹, Mariana Callil Voos², Isabela Pessa Anequini Leite², Francis Meire Favero¹, Acary Souza Bulle Oliveira¹, Fátima Aparecida Caromano²

1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

Introduction. The girdle muscular dystrophy type 2A (calpainopathy) is a neuromuscular disease with autosomal recessive inheritance, clinically characterized by muscle weakness and atrophy and progressive symmetrical limb and pelvic girdle muscle weakness. Few studies characterize muscle force in different body segments in patients with calpainopathy. **Objective.** To characterize the muscle force in patients with calpainopathy in a Brazilian sample. **Method.** Retrospective, cross-sectional, and descriptive study conducted at Laboratory of Physiotherapy and Behavior, Medical School and Center for Human Genome and Stem-cell Researches, both from the University of São Paulo, and Department of Neurology of the Federal University of São Paulo. We studied thirty-three historical of patients evaluated from December 2008 to June 2014, all with molecular diagnostics (DNA or muscle biopsy) of calpainopathy. We used the data of muscle force, collected using manual muscle test of the groups flexor, extensor and abductor of the shoulder; flexor and extensor of the elbow, wrist, knee and hip; dorsal and plantar flexors of the foot. For the characterization of the patients (age and sex) and the muscle force test descriptive statistical analysis was used. **Results.** We found weakness with proximal predominance of upper and lower limbs in all 33 patients. **Conclusion.** It was possible to characterize the muscle force of different muscle groups of Brazilian sample of patients with calpainopathy, reinforcing that muscle weakness in people with this disease is predominantly proximal.

Keywords. Girdle Muscular Dystrophy, Calpainopathy, Muscle strength.

Clinical Characteristics in the Autosomal Recessive Limb-Girdle Muscular Dystrophy - Sarcoglycanopathy: A Report of Cases

Geisa Silva Cunha Moraes¹, Isabela Pessa Anequini Leite², Francis Meire Favero¹, Michele Emy Hukuda², Acary Souza Bulle Oliveira¹, Mariana Callil Voos², Fátima Aparecida Caromano²

1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

Objective. Characterize the standard of muscle weakness and clinical features of patients with autosomal recessive limb-girdle muscular dystrophy - sarcoglycanopathy type. **Method.** Descriptive analysis using retrospective banks of the Centro de Pesquisa over the Genoma Humano and Células Tronco, USP with 18 individuals diagnosed with sarcoglycanopathies. **Results.** There was a mean age of 21.4 years, ranging between 8 and 47 and the duration of illness was 14.5 years. The prevalence was women and wheelchair users, 67% respectively. Of these eighteen individuals, three individuals with DMC2C, six with DMC2D, two DMC2E, two DMC2F and five individuals with DMC sarcoglycan with multiple proteins disabilities. The onset of symptoms varied from childhood to early adolescence. The gait loss was also in adolescence. There are also cases of adults who still walk and more cases of late-onset disease. The DMC2D was the group more homogeneous with half of the subjects with calves hypertrophy, all subjects featuring scoliosis and contractures of the lower limbs in 67% of cases. **Conclusions.** The study identified the onset of weakness muscle in the proximal muscles, with a predominance of pelvic girdle associated with other changes, but more studies are needed to understand the genetic mutations with different phenotypes and thus propose more effective physical therapy interventions. However, more studies are needed to understand the genetic mutations with different phenotypes. And so, propose more effective physical therapy interventions.

Keywords. Girdle Muscular Dystrophy, Sarcoglycan, Physical Exam, Weakness Muscular, Disorders of Motor Abilities.

Literature Review on the Assessment Tools that showed Responsiveness in Neuromuscular Diseases

Tatiana Aparecida dos Santos Veríssimo¹, Michele Emy Hukuda², Francis Meire Favero¹, Acary Souza Bulle Oliveira¹, Mariana Callil Voos², Fátima Aparecida Caromano²

1.Universidade Federal de São Paulo (UNIFESP), São Paulo-SP, Brasil.

2.Universidade de São Paulo (USP), São Paulo-SP, Brasil.

Objective. To identify which assessment tools have responsiveness in patients with neuromuscular diseases and different focuses of the research on the responsiveness of the assessment instruments in neuromuscular diseases.

Method. This is a bibliographic review on the subject "responsiveness of the assessment instruments in the neuromuscular disorders", from January 2004 to December 2014. The studies were obtained from the Latin American database and Caribbean Health Sciences, the National Library of Medicine, and the Scientific Electronic Library Online. For inclusion in the study were read in full, since the responsiveness psychometric property may not have been described in the summary. Studies were selected by two physical therapists with specialist training and master. After reading and the inclusion of studies in this research, the tests used to assess the responsiveness and identify the focus of analysis of the responsiveness of each instrument have been described. **Results.** It was found the existence of twenty-six studies. Of these six, they presented responsiveness. The instruments that had their responsiveness tested in patients with neuromuscular disorders were ACTIVLIM, Motor Function Measure, Functional Independence Measure, Barthel Index, Rehabilitation Activities Profile and North Star Ambulatory Assessment. **Conclusion.** The literature review showed that the number of studies that tested the responsiveness of a measuring instrument in the neuromuscular disorders was scarce, only in six evaluation tools. Several research focus were found to analyze a range of responsiveness, because there is still no gold standard. Thus, recognizing the validity of a measuring instrument is not definitely proven, but sustained the evidence accumulation, there is the need for further studies to evaluate the psychometric properties, especially the responsiveness.

Keywords. Neuromuscular diseases, Questionnaires, Sensitivity and Specificity, Evaluation, Task Performance and Analysis.

