Theranostics of
Brain, Spine & Neural Disorders

Juniper Publishers
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Research Article
Volume 3 Issue 5 - September 2018

Theranostics Brain Disord

Physiotherapy Intervention in Delayed Motor Evolution in a Clinical Case of Neuromuscular Disease

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Submission: August 31, 2018; **Published:** September 20, 2018

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Abstract

Child development is a process that begins in intrauterine life. These abilities are developed in the typical developmental child, may generate deviations from this development, being related to some pathology, which generates the atypical development of the child. Methodology: The aim of this study was to demonstrate the performance of the physiotherapy with application of the Bo bath concept Campos, Santos, Tudella, Pereira & Rocha in the evolution delay in a clinical case of neuromuscular disease. Data collection from February 2008 to February 2014 from the clinical analysis and the application of the Gross Motor Function Measure (GMFM) and of the Schedule of Growing Skills II (SGSII) in a three-year period. Results: The evolution of the disease showed a severe loss of the capacities evaluated by the test of SGSII. In the initial evaluation according to the GMFM the patient presented a global result of 39.6% and at the end of the study, the patient did not perform active movements, obtaining a total value of 0%. Conclusion: The intervention of Physiotherapy in this clinical case was an advantage in the maintenance of voluntary movement patterns and joint amplitudes, as well as in the reduction of the rapid evolution of motor losses and what our role as professionals in providing help and guidance to users, families and the surrounding community

Keywords: Physiotherapy Intervention;Bo bath Concept; Neuromuscular Disease; Gross Motor Function Measure; Schedule of Growing Skills Ii

Introduction

Child development is a process that begins in intrauterine life. At the moment of fertilization[1], the newly formed human being carries a somatic and psychic contingent corresponding to his species, his race and his ancestors Burns et al. [2],Rotta [3]. When we look at the motor component of child development, Goldberg et al.[4] report that motor development is the process of change in motor behavior, which is related to age, both in the posture and in the movement of the child. Throughout your publications, several authors have demonstrated the need to study child development and its deviations in different age groups, due to the innumerable factors that can influence child acquisition and performance over the years Gallahue et al. [5], Papalia et al. [6],Papalia et al. [7], Payne et al. [8].These abilities are developed in the typical developmental child Goldberg et al. [4],Papalia et al. [7], FortiBellani et al.[9] and when there is any alteration and/or development, may generate deviations from this same development, being related to some pathology, which generates the atypical development of the child Forti-Bellaniet al. [9],Flehming [10],Bezerra et al.[11], neuromuscular disorder is one of the most common causes of this atypical development (Arenas &Gispert)Correia [1],Emery[12], Brice[13]. Neuromuscular disease (NMD), is a generic term, which affects one in 3.500 children worldwide, which are grouped different diseases resulting from diseases affecting the motor unit, consisting of the spinal motor neurons, nerve root, peripheral nerve, myoneural junction and muscle Emery [12],Reed[14],Féasson et al. [15-17],Dinsmore[18].

One of the examples of neuromuscular disease is infantile neuroaxonal dystrophy. Spielberg's syndrome, also known as infantile neuroaxonal dystrophy, is a rare, neurodegenerative pathology with autosomal recessive inheritance Scola et al. [19]. Clinical manifestations of the disease usually occurs between six month and 3 years old. The initial symptom is a slowing of the rate of motor and mental development followed by a early visual changes such as decreased visual acuity and optic atrophy, leading to blindness, hyperreflexia, atrophy, hypotonia and muscular weakness and, finally, dementia. Seizures are rare, but when present, may indicate a terminal condition of the Syndrome Scola et al. [19], Brice[13],Perry &Scheithauer[20],Nardocci [21].The intervention of neuromuscular disease and in specific of this type of pathology is based on the constantstimulation of the child, taking into account that the maintenance of the state of the same is a therapeutic gain. The ultimate goal is to provide tools for a higher quality of life and less suffering for the child, such as facilitating the functions of their daily caregivers Brice[13],Lui &Byl[22],Fell[23].Thus, the analysis and follow-up of a clinical case of neuromuscular disease, more specifically a clinical case of infantile neuroaxonal dystrophy, over a

long period of time may allow a more holistic approach in the search for improvement in the quality of life of the child and caregivers, structuring the environment in order to minimize the losses of the child's motor capacities.

Methodology

Aim: This article aimed to present a study on the performance of physiotherapy with application of the Bobath concept Campos et al.[1] in the delay of the evolution of a clinical case of neuromuscular disease, especially in a clinical case of Spielberg's syndrome or Neuroaxonal Infantile Dystrophy (IND) and its influence in the motor development of the child.

Study Sample: The source and data collection were carried out by the principal investigator from February 2008 to February 2014. Data analysis was performed correlating the evolution of the disease with the treatment performed and its interference in the clinical picture of a patient.The case study is one of the oldest methods used in teaching and can be used in continuing education Galdeano et al.[24]. The Case Study Method fits as a qualitative approach and is often used for data collection Denzin& Lincoln[25].Approved by the ethical comity of Piaget Institute, this study asked for informed consents for students' parents and guardians. This project is in line with national and international guidelines for scientific research involving human subjects, including the Declaration of Helsinki in 2013 on ethical principles of medical research in human beings and the 1997 Convention on Human Rights and Biomedicine (the "Oviedo Convention").

The inclusion criteria are:

a) To be aged between 10 and 11 years;

b) Understand the aims of the study and agree to participate voluntarily.

As exclusion criteria:

a) Presence of pathology a known health condition of rheumatological, orthopedic, cardio-respiratory, oncological pathology and/or diagnosed coronary disease;

b) The provision of informed consent by students' parents and guardians

Procedures: The use of a single case is appropriate in some circumstances: when the case is used to determine whether the propositions of a theory are correct; When the case under study is rare or extreme, that is, there are not many similar situations for comparative studies Stake [25].

In this sense, a case report of a female child with Spielberg's Syndrome (Infantile Neuroaxonal Dystrophy) was carried out. The total duration of the study was six years (between the 2 years and 9 years of age of the child) with the period of bi-weekly therapeutic intervention. The child was in the ambulatory service of the Portuguese Association of Cerebral Palsy of Leiria.The clinical history of the patient, the main complications according to clinical evolution, the proposals for treatment by physical therapy and the therapeutic response of the patient were described in this study. The source and data collection were carried out by the principal investigator from February 2008 to February 2014, using data from the clinical analysis and the application of the Gross Motor Function Measure (GMFM) and Development of the Schedule of Growing Skills II in a three-year period.Data analysis was performed correlating the evolution of the disease with the treatment performed and its interference in the clinical picture of a patient. Due to the degenerative nature of neuromuscular affections, there is currently no treatment other than palliative and preventive Lima et al.[26]. In this way, physiotherapy works together with the multidisciplinary team in the prevention and treatment of signs and symptoms of the disease, thus contributing to a greater and better survival of the patients.For the statistical analysis of the results, descriptive statistics were used using the standard methods, using the statistical treatment program Statistical Package for the Social Sciences (SPSS), version 21.0.

Clinical Case Study of Infantile Neuroaxonal Dystrophy

Child A.J.A.S., was born on 10 September 2005 of a watched pregnancy, with delivery at 40 weeks by cesarean section, with somatometry at birth: weight 3810 g; Stature 50 cm; Cephalic perimeter 35cm; Apgar index 9/10. No relevant perinatal history and good adaptation to extrauterine life.As for the development of the child, he smiled at the first month, produced the first syllables at 5 months and the first words at 18 months. He held his head at 4 months, sat at 8 months, crawled at 14 months, marched with support at 18 months.The approach of Physiotherapy in neuromuscular diseases does not differ in specific to the clinical cases of Infantile Neuroaxonal Dystrophy (IND). The establishment of therapeutic activities and/or exercises for patients with neuromuscular diseases should take into account the natural course of the disease, as well as the effects of exercise on the organic system and the molecular adaptations observed in the short and long term. Therapeutic exercises should be seen as a way to improve the quality of life or to minimize the loss of motor capacity that occurs in neuromuscular diseases in general and more specifically in IND, and the objectives are directed to an improvement in the performance of related activities of the child Orsini et al. [27] and Fowler [28].

A.J.A.S. Began Physical Therapy in February 2008. He was totally dependent on the activities of daily life but always encouraged his collaboration in activities, he performed active movements of the upper and lower limbs.In the evaluation by direct spontaneous/provoked observation, a girl attentive, but not always cooperative, trying to accomplish only that which has will and having behaviors of rejection when this does not happen. He would sit and hold the position without support from the upper limbs on the mattress or on the low bench, he would not crawl, but he would drag himself forward in the sitting position with his hip, and he had some difficulties in transferring from sitting down from the bench to the floor and vice She stood up with support, but she did not stand on her own. She was walking with two-hand support.To enhance their autonomy in the day-to-day, the strategies were to improve

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the postural control, stimulate the exploration and manipulation of objects/toys and sensory stimulation.The approach of Physiotherapy in neuropediatric is to encourage the development of the child through games appropriate to his age and oriented to functionality.

The goal of physical therapy in neuropediatric is to minimize the effects of disability, promote function and musculoskeletal development. The therapeutic approach is holistic and practical, with greater emphasis on motor function and posture Lima et al.[26],Moreira et al.[29],Tecklin[30],Desouzart et al. [31].The methods and techniques developed to achieve the proposed goals were the Neurodevelopment Techniques (NDT) Raine et al.[32] and the Proprioceptive Neuromuscular Facilitation (PNF) techniques Moreira et al. [29].A.J.A.S. Evolved to major limitations of active and passive movement, developing several joint deformities: flexion of the elbows, hips and knees; (Left and right), right hypotonia, and hypertonia of the upper and lower limbs, and which meet several studies Scola et al. [19],Brice[13]. At the end of 2010, the user no longer performed active anti-gravitational movements.At the end of the study (February 2014), the patient did not perform active movements, not being able to perform any activity, does not control the head or the trunk, presents spasticity of the lower and upper limbs and has great joint limitations, especially at the level of the feet.The rapid loss of their capacities led to the reformulation of the objectives. These focus on normalization of muscle tone and maintenance of range of motion (avoid further muscle retraction and deformities) through passive mobilization and postural postures through NDT techniques and sensory stimulation Moreira et al.[29]Raine et al.[32], to provide comfort and well-being for the child and structural mechanisms for their caregivers to handle and care for the child Correia [33].

Resultsand Discussion

A.J.A.S. Was assessed with the Schedule of Growing Skills II Developmental Test (Lingam, Hunt, Golding, Longmans, & Emond) for 24 months: Active posture: 10 months; Locomotion and Nonverbal Cognition: 12 months; Manipulation, Vision, Speech/Language and Autonomy: 15 months e; Listening/Language and Social Interaction: 18 months Table 1.At 36 months, it was re-evaluated with the Schedule of Growing Skills II Development Assessment Test, where 1 year later, development levels in the evaluated areas practically remained, with some areas declining. This presentation 1 year after the first evaluation, allowed to verify the beginning of the degenerative process.At 60 months, the evolution of the disease showed a severe loss of the capacities evaluated by the test, and the parameters evaluated did not allow to obtain quantifiable data to fill the test. The same happened at 84 months, where we did not obtain measurable data. These comparative results are shown in Table 1.In the Gross Motor Function Measure (GMFM) assessment, the patient was evaluated triennially (beginning, middle and end of the proposed intervention program).In the initial evaluation according to the GMFM in December 2007, at 2 years and 2 months (26 months), the patient presented a global result of 45.9% of the gross motor capacity, having obtained 100% in dimension A (decubitus and rolling) , 71.7% in dimension B (sitting posture), 26.2% in dimension C (cats and knees), 15.4% in dimensions D (standing posture) and 12.5% in dimension E (walking, running And jump).

Table 1: Schedule of Growing Skills - II test results from 2007 to 2014.

Age of assessment	24 month (2007)	36 month	60 month	84 month
Areas evaluated		-2008	-2011	-2014
Passive Posture		3		
Active Posture	10	6		
Speech/Language	15	12		
Non-verbalcognition	12	12		
Manipulation	15	15		
Autonomy	15	15		
Hearing/ Speech	18	18		
Social interaction	18	18		
Vision	15	18	0	
Locomotion	12			

In February 2008, at 2 years and 5 months (29 months), in only 3 months after the first evaluation, the patient presented a regression of the capacities with an overall result of 39.6% of the gross motor capacity, obtaining 100% in the Dimension A, 71.7% in dimension B, 26.2% in dimension C and 0% in dimensions D and E.In February 2011, at 6 years and 5 months, the patient was reassessed according to GMFM and presented a total value of 0.51 of the gross motor capacity, obtaining 3.9% in dimension A and 0% B, C, D and E.At the end of the study (February 2014), at 8 years and 5 months, the patient did not perform active movements, being unable to perform any activity proposed for the GMFM, obtaining a total value of 0% of gross motor capacity. The comparison of overall and specific results are presented in Figure 1.The presentation of these final data allows a comprehensive analysis of the great loss of functional capacity, which leads to a readaptation of the child's and his / her caregivers' lifestyle, with a rapid regression of their motor skills, generating a progressive dependence in all areas Life (health, social and environmental)[33-37].

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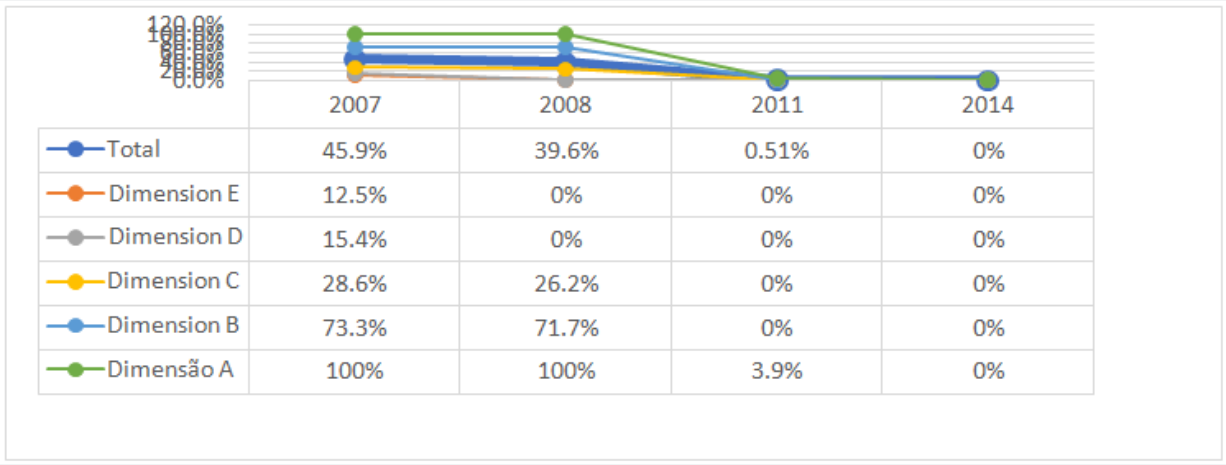

	2007	2008	2011	2014
Total	45.9%	39.6%	0.51%	0%
Dimension E	12.5%	0%	0%	0%
Dimension D	15.4%	0%	0%	0%
Dimension C	28.6%	26.2%	0%	0%
Dimension B	73.3%	71.7%	0%	0%
Dimensão A	100%	100%	3.9%	0%

Figure 1: Graph 1-Triennial evolution of motor skills according to GMFM from 2008 to 2014.

Conclusion

Facing the problem of neuromuscular diseases, which occurs in all current societies, have been developing a concern and a new way of facing the clinical process of each user Correia.The goal of Physical Therapy in the field of neuropediatric is to help children reach their full potential for functional independence through analysis, assessment, health promotion and wellness, and through the implementation of a wide variety of interventions and supports. The Physiotherapist's role is to assess the overall development of the child as well as specific physical difficulties, and to provide users, parents and community with guidance regarding the handling, positioning and treatment of the user through specific games, exercises and / or functional activities.It also has an important role in the choice and monitoring of support products that facilitate movement and help maintain independence, such as orthoses, gaiters, wheelchairs and positioning devices, among others.

The longevity gained by the progress of science and the improvement of the quality of life promotes the opportunity of the neuromuscular patient to enjoy a longer life, compared to the time of survival, and stimulates the development of preventive methods and techniques with regard to Maintenance of movement and posture patterns, reducing deformities and avoiding degradation of the health condition.

We can conclude that the intervention of Physiotherapy in this clinical case of neuromuscular disease was an advantage in the maintenance of voluntary movement patterns and joint amplitudes, as well as in the reduction of the rapid evolution of motor losses and this goes according to the indications of several authors (Orsini, et al.,Strehle, Arenas &Gispert, Fell). In aims terms, the realization of this case study in the construction of a long-term study provided the notion of how much we need to be aware of this problem, and what our role as professionals in providing help and guidance to users, families and the surrounding community. Given this statement, the proposal for continuity of the study in a more comprehensive way and the awakening to this theme remains.

Acknowledgement

The author would like to thank the parents of the child A.J.A.S., Rui and Alice, fighters who deserve all the recognition and affection and the Portuguese Association of Cerebral Palsy of Leiria.

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