

Aquatic Physical therapy in a patient with Limb-Girdle Muscular Dystrophy, type 2b: a case report

Fisioterapia aquática em paciente com distrofia muscular de cinturas do tipo 2b: relato de caso

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ABSTRACT | INTRODUCTION: Limb-Girdle Muscular Dystrophy, Type 2B (LGMD2B), is a rare, hereditary, progressive neuromuscular degenerative disease coursing with progressive impairments in motor and functional capacity. **OBJECTIVE:** To describe and analyze the effects of aquatic physical therapy on the functionality, muscle strength, range of motion, and quality of life of a patient diagnosed with LGMD2B attended on an outreach program. **METHODS:** A female patient, 32 years old, single, with genetic diagnosis of LGMD2B, level 5 at Vignos scale (modified by Garder-Medwin e Walton). The case reports the Aquatic Physical Therapy rehabilitation protocol (hydrokinesiotherapy) and its impacts on muscle strength, range of motion, functional capacity, and patient quality of life (CAAE No. 43505321.0.0000.0018). **RESULTS:** The aquatic physical therapy protocol, composed of 12 sessions, 60 minutes/2x/week, resulted in improvements in overall functional capacity and a 9.52% increase of distal motor function, 100% increase in handgrip strength, and increase up to the upper limit (grade 5) on the MRC scale for several of the muscles tested, in addition to increased range of motion and expressive improvement in Quality of Life. **CONCLUSION:** The patient' functional improvement suggests that water-based physical therapy rehabilitation, at mild to moderate exercise intensity, is a safe and effective therapeutic option for improvement muscle strength, range of motion, functional capacity, and quality of life in LGMD2B patients.

KEYWORDS: Hydrotherapy. Muscular Dystrophies, Limb-Girdle. Muscular Dystrophies. Rehabilitation. Physical Therapy Specialty. Case Reports.

RESUMO | INTRODUÇÃO: A distrofia muscular de cinturas do tipo 2B (DMC2B) é uma doença neuromuscular, degenerativa, rara, hereditária, progressiva, com consequentes prejuízos progressivos na capacidade motora e funcional. **OBJETIVO:** descrever e analisar os efeitos da fisioterapia aquática sobre a funcionalidade, força muscular, amplitude de movimento e qualidade de vida de uma paciente com diagnóstico DMC2B atendida em projeto de extensão universitária. **MÉTODOS:** Paciente do sexo feminino, 32 anos, solteira, com diagnóstico genético de Distrofia Muscular de Cinturas do Tipo 2B, nível 3 da escala Vignos (modificada por Garder-Medwin e Walton). O relato de caso apresenta a reabilitação através da Fisioterapia Aquática (hidrocinesioterapia) e seus impactos sobre a força muscular, amplitude de movimento, capacidade funcional e qualidade de vida da paciente (CAAE No. 43505321.0.0000.0018). **RESULTADOS:** O protocolo de fisioterapia aquática, composto por 12 sessões, 60 min/2x/semana, resultou em melhoras na capacidade funcional global e aumento de 9,52% na avaliação da função motora distal, aumentos de 100% da força de preensão manual e aumento para o limite superior (grau 5) na escala MRC para várias das musculaturas testadas, além de ganho de ADM e melhora expressiva da Qualidade de Vida. **CONCLUSÃO:** A melhora funcional apresentada pela paciente sugere que a reabilitação funcional fundamentada na fisioterapia aquática, em intensidade leve a moderada, é uma opção terapêutica segura e eficaz para a melhora da força muscular, amplitude de movimento, capacidade funcional e qualidade de vida na DMC2B.

PALAVRAS-CHAVE: Hidroterapia. Distrofia Muscular do Cíngulo dos Membros. Distrofias Musculares. Reabilitação. Fisioterapia. Relatos de Casos.

Introduction

Limb-girdle muscular dystrophies (LGMD) are a group of progressive, hereditary, rare, and heterogeneous neuromuscular diseases^{1,2}, with symptoms mainly related to the impairment of the muscles of the pelvic and scapular girdle and proximal extremity.³ LGMD estimated prevalence ranges from 1 in 14,000 to 45,000 individuals and may vary according to specific subtypes and populations.² Limb-girdle muscular dystrophy, type 2B (LGMD2B), is the second most frequent subtype in many countries (15 to 25%). A Brazilian study indicated a prevalence of 9.79% for LGMD, with 18% corresponding to LGMD2B.⁴

In LGMD2B, an autosomal-recessive mutation occurs in the DYSF gene, which encodes the dysferlin protein in skeletal muscle cells membrane, resulting in muscle weakness, mainly on the proximal muscles.⁵ Recent magnetic resonance imaging (MRI) studies demonstrate lower limb muscles such as the gluteus, tensor fasciae latae, semitendinosus, semimembranosus, biceps femoris, and triceps surae are severely affected.²

Range of motion, mobility, muscle function, and quality of life are progressively affected by LGMD2B, impairing functionality, and leading to occupational, social, and economic losses.⁶ Cardiac or respiratory impairments are uncommon, clinically silent, and may be present later in life.⁷

There is no cure for LGMD. Current treatment techniques include gene and drug therapy, surgical intervention, support for cardiac and respiratory functions, emotional and physical support, in addition to assistive devices such as canes, walkers, and wheelchairs.⁸ Physical therapy has an important role in preserving and improving the LGMD2B patient's quality of life, aiming maximum independence with emphasis on functional and muscle-strengthening activities.⁹

A recent review pointed out that there is not enough high-level evidence to support the specific recommendation for aquatic physical therapy inclusion in treating muscular dystrophies patients.¹⁰ Findings of the review related to limb-girdle muscular dystrophy, the object of interest in this manuscript, are limited to a single case study.¹¹ It is also essential to highlight the need for aquatic physical therapy intervention protocols registration, in association

with the description of the patient's functional status, to support its replication in clinical practice, justifying the need of this case report, derived from observations and intervention carried out in a clinical setting.¹⁰

In this context, this study aims to describe and analyze the effects of aquatic physical therapy protocol, associated with ground home exercises orientations, on functionality, muscle strength, range of motion, and quality of life of a patient with a genetic diagnosis of LGMD2B treated in a university outreach program.

Case Report

Clinical case presentation

We present a case report of a 32-year-old female patient, single, right-handed, sedentary, with no history of sports practices or functional rehabilitation, with a genetic diagnosis of limb-girdle muscular dystrophy, type 2B, confirmed by a medical report attested by a geneticist. The patient was classified as level 3 of the Vignos scale (modified by Garder-Medwin and Walton).¹²

The patient, whose parents are first cousins, reported four relatives (one brother, two sisters, and one nephew) genetically diagnosed for LGMD2B. In addition, she also reports an aunt and a cousin with the same clinical condition and similar evolutions. Unfortunately, the research team did not have access to clinical or diagnostic information of family members.

The patient reported symptoms onset in 2008 (eight years before joining the outreach program), including loss of balance and lower limbs muscle weakness, without activities limitations. The clinical condition progressed slowly to activities limitations of climbing stairs and kneeling in 2011, and exacerbating in 2014, when she started to perform those activities only when extremely necessary, choosing to use preferably ramps.

Pre-intervention kinetic-functional assessment in 2016 showed predominantly proximal paraparesis, postural instability, and patient report of hips and knees joint pain and fatigue on medium efforts. The patient performed activities independently, with

limitations, and the need of upper limb assistance to perform changes in basic body position, e.g., changes in decubitus, sitting and standing, or transferring from a kneeling to a standing position. The patient was able to stand independently, with occasional need for upper limb support, maintaining an unstable upright posture, predominantly using an enlarged base of support. She presented a waddling gait and difficulties for walking medium distances, like decreased speed and cardiorespiratory fatigue. The patient maintained functionality to go up and downstairs, requiring handrail support and a longer time to complete the task.

The above activities limitations led to a reduced group and social participation, especially those associated with a nun routine tasks, such as transfer from the kneeling to standing position and the limitations of group walking transfers due to the decrease in gait speed.

The patient reported the chief complaints: pain in the left knee joint, fatigue on medium efforts, and limitation in activities of daily living (ADLs), which guided the rehabilitation protocol. This study followed the recommendations of Resolution number 466/2012 and the Declaration of Helsinki and was approved by the Institutional Ethics Committee (CAAE No. 43505321.0.0000.0018). The patient signed the Informed Consent Form.

Intervention Protocol

The intervention program was composed of twelve sessions (60 min/2x/week) of aquatic physical therapy during six weeks to treat reduced muscle strength, knee pain, and impairment of functionality in ADLs. Each session included: stretching, functional training, cardiorespiratory conditioning, and muscle strengthening (Table 1), associated with orientations of home-ground exercises for fine motor coordination. The patient did not undergo any other physical treatment concomitantly with the aquatic physical therapy.

Table 1. Aquatic physical therapy intervention program protocol

Stretching	Cardiorespiratory Training (Global Training)	Muscle Strengthening*
1. Shoulder extensors and flexors muscles; 2. Hip extensors and flexors muscles; 3. Knee extensors and flexors muscles; 4. Ankle plantar flexors and dorsiflexors muscles; 5. Girdles dissociation on flotation*.	1. Awareness exercises of breathing pattern; 2. Bicycle movements on flotation*; 3. Walking back and forth (long steps and frontal turbulence); 4. Swimming movements (front and back crawl, and breaststroke) #	1. Shoulder extensors, flexors, adductors, and abductors muscles; 2. Elbow extensors and flexors muscles; 3. Wrist extensors and flexors muscles; 4. Abdominal muscles; 5. Hip extensors, flexors, adductors, and abductors muscles; 6. Knee extensors and flexors muscles; 7. Ankle plantar flexors and dorsiflexors muscles.

(*) Floating pool noodles foam tubes, flutter boards, water dumbbells, and aquatic fingerless gloves were used according to the need for support or resistance for each movement.

(#) The various swimming movements were performed according to the patient's potential and possibilities in an aquatic environment.

Global stretching for upper and lower limbs major muscle groups (progressing from static to dynamic) or passive stretching in fluctuation were used at the beginning and the end of each session. The cardiorespiratory training was performed using walking exercises against turbulence, swimming patterns, or ride-in bike simulated movements on flotation and had an average duration of 30 minutes on intensity equivalent to level 5 on the Borg scale (mild to moderate intensity). The exercises for muscle strengthening were carried out against water resistance, using materials such as flutter boards, water dumbbells, floats, and aquatics gloves, with an average duration of 20 minutes. Exercises were associated with manual stimulation (sliding Tapping) as tactile feedback to enhance muscle contraction, performed by the therapists.

The difficulty progression to increase resistance to the movement was performed by manipulating variables: speed and direction of movement, shape, position, depth of equipment, and/or turbulence. The patient was also instructed to perform ground exercises, focusing on fine motor coordination of hands and fingers, using easily accessible materials, such as small stones and cooking grains.

Assessment

The same researcher performed the pre-and post-interventions assessments, including clinical history, functional capacity (Motor Function Measure scale - MFM), muscle strength (Medical Research Council Scale - MRC and manual dynamometry), range of motion (manual goniometry), and quality of life (SF-36). All tests are widely used in scientific and clinical settings to assess patients with neuromuscular diseases.¹³⁻¹⁷

Functional capacity

The Motor Function Measure (MFM) scale is specific and sensitive for the functional assessment of patients with neuromuscular diseases.¹⁸ This scale comprises 32 items divided into three dimensions (D1, D2, D3) being designed to provide a detailed profile of the motor function of the standing position and transfer (D1), the axial and proximal motor function (D2), and distal motor function (D3). The total score ranges from 0 to 96 points, corresponding to low and high motor

function, respectively, also possible to calculate each dimension's value as a percentage.¹⁵

Muscle strength

Muscle strength assessment was performed manually (MRC scale) and by handgrip strength measurement (dynamometry).^{13,14,19} The MRC score ranges from '0' to '5', where '0' represents no visible muscle response, '1' visible or palpable muscle contraction, without limb movement, '2' limb movement but not against gravity, '3' movement against gravity throughout the almost complete range of motion, '4' movement against moderate resistance throughout the range of motion, and '5' normal movement. Muscle strength was assessed bilaterally in Brachial biceps, brachial triceps, deltoids, wrist flexors and extensors, shoulder adductors, quadriceps femoris, iliopsoas, hamstrings, hip abductors and adductors, gastrocnemius, tibialis anterior, gluteus medius, and maximum.

Handgrip strength has been assessed using a hydraulic dynamometer (Jamar®), following the positioning and data collection recommendations of the American Society of Hand Therapists.¹⁹ Three attempts were made, with intervals of 30 seconds between measurements. The best performance was used for analysis.

Range of Motion (ROM)

The assessment of ROM was performed by manual goniometry (goniometer Carci®), a universal method of use, with advantages including low cost and easy-to-use instrument. ROM was assessed at the end of the full range of motion, bilaterally, flexion, extension, abduction, and adduction of shoulders and hips, flexion and extension of elbows, wrist, knees, plantar flexion, and dorsiflexion of the ankles.

Quality of Life

Quality of life was assessed using the SF-36 questionnaire (Short Form Health Survey 36), an instrument composed of 36 items, widely used for assessment of the health-related quality of life, including with muscular dystrophies population.^{16,17} The SF-36 questionnaire assesses eight health domains: physical functioning, function limitations

due to physical health problems, bodily pain, general health, vitality, social functioning, function limitations due to emotional problems, and mental health. The total score ranges from 0 (zero) to 100 (one hundred) points, with higher scores indicating better general health.

Results

At the pre-intervention assessment, the patient was in good general health. However, it was observed: independent and waddling gait and weakness in the upper and lower limbs.

In the assessment of MFM scale after 12 sessions of aquatic physical therapy, there was a 6.25% increased in the functional capacity total score, with 7.69% improvement on motor function for the standing position and transfers, 2.78% on axial and proximal motor function, and 9.52% on the distal motor function (Table 2).

Table 2. Functional capacity. Scores for Motor Function Measure Scale (MFM) dimensions at pre- and post-interventions assessments. Improvement percentage for each dimension is also shown

MFM scale dimensions	Pre-Intervention (%)	Post-Intervention (%)	Improvement (%)
D1	66.7	74.36	7.69
D2	97.22	100.00	2.78
D3	85.71	95.24	9.52
Total	82.29	88.54	6.25

%; percentage. D1: Dimension 1 - Motor function of standing position and transfers; D2: Dimension 2 - Axial and proximal motor function; D3: Dimension 3 - Distal motor function.

Pre-intervention assessment of muscle strength by using the MRC scale indicated impairment in both the distal and proximal muscles, with more significant impairment of lower limb muscle strength (MRC scores ≤ 4) than in the upper limbs (MRC scores ≥ 4) (Table 03). After aquatic physical therapy intervention, an increase of 1 degree of strength in wrist flexors and extensors, shoulder adductors, biceps brachial, deltoids, and maintenance of the strength measured in the brachial triceps were observed. At post-intervention assessment, all muscle groups in upper limbs, which were tested as grade 4 at the initial assessment, improved to grade 5, the maximum strength score (Table 3).

In the lower limbs, quadriceps femoris strength was maintained. Gastrocnemius, tibialis anterior, gluteus medius, gluteus maximus, hamstrings, hip adductors, and iliopsoas muscles strength increased 1 degree. Hip abductor muscle strength showed an increase of 2 degrees, reaching grade 4 at post-intervention assessment (Table 3).

Table 3. Muscle strength assessment - Medical Research Council (MRC) scale. Results are presented in absolute values for the pre- and post-intervention assessments

Muscles assessed	Right Upper Limb		Left Upper Limb	
	Pre-Intervention	Post-Intervention	Pre-Intervention	Post-Intervention
Brachial Biceps	5	5	4	5
Brachial Triceps	5	5	5	5
Deltoids	5	5	4	5
Wrist Flexors	4	5	4	5
Wrist Extensors	4	5	4	5
Shoulder Adductors	4	5	4	5
Muscles assessed	Right Lower Limb		Left Lower Limb	
	Pre-Intervention	Post-Intervention	Pre-Intervention	Post-Intervention
Quadriceps Femoris	4	4	4	4
Iliopsoas	4	5	4	5
Hamstrings	3	4	3	4
Hip abductors	2	4	2	4
Gastrocnemius	1	2	1	2
Hip adductors	4	5	4	5
Tibialis Anterior	1	2	1	1
Gluteus Maximus	4	5	4	4
Gluteus Medium	1	2	1	2

The handgrip assessment indicated an increase of 53.3% and 100% in handgrip strength after the intervention for the right (pre-intervention: 7.5 KgF, post-intervention: 11.5 KgF) and left upper limb (pre-intervention: 5 kgF, post-intervention: 10 kgF), respectively. Range of motion (ROM) assessment of the main joints of the upper and lower limbs increased after the aquatic intervention protocol for all joints tested. It is important to highlight no mechanical limitations or joint stiffness to movements at pre-intervention assessment.

The greatest improvements on ROM were observed for shoulder flexion (Right: + 42°; Left: + 43°), shoulder abduction (Right: + 33°; Left: + 21°), elbow flexion (Right: + 26°; Left: + 24°), knee flexion (Right: + 13°; Left: + 7°), knee extension (Right: + 10°; Left: + 19°), hip abduction (Right: + 10°; Left: + 6°), in addition to plantar flexion (Right: 1°; Left: 4°).

Quality of life, assessed by the SF-36, improved after aquatic physical therapy intervention. The patient showed greater improvements in physical (400%), pain (80%), and functional capacity (37.5%) domains. All other domains had sustained or increased scores after rehabilitation, except the mental health domain, as shown in Table 4.

Table 4. Quality of Life Assessment (SF-36). Results are presented in absolute values for pre-and post-intervention assessments. The percentage of change between assessments for each dimension is also shown

Domains	Pre-Intervention	Post-Intervention	Difference (%)
Functional capacity	40	55	+37.5
Limitations due to physical aspects	25	100	+400
General health status	40	47	+17.5
Vitality	65	75	+15.38
Limitations due to emotional aspects	100	100	0
Pain	40	72	+80
Mental health	88	72	-18.18
Social aspects	100	100	0

%; percentage.; +: gain; -: loss.

Discussion

This study aimed to analyze the effect of aquatic physical therapy intervention protocol on functionality, muscle strength, range of motion, and quality of life of limb-girdle muscular dystrophy, type 2B (LGMD2B) patients. Our results show that the protocol of aquatic physical therapy applied during 12 sessions improved the overall functional capacity and increased the distal motor function by 9.52%. Also increased the left handgrip strength by 100%, and the MRC scale score for all muscles with grade 4 at the assessment, reaching the higher score (grade 5). In addition, ROM and quality of life improved.

A physically active lifestyle and the practice of physical exercise for health promotion, prevention, and treatment of different diseases is an international recommendation. However, the recommendation for intervention through physical exercise is controversial in people with neuromuscular diseases, although there is evidence of its benefits as a treatment for this group of patients.^{14,20,21}

As a rare disease resulting from a specific recessive genetic mutation, studies investigating specific functional rehabilitation strategies for people affected by limb-girdle muscular dystrophy type 2B(LGMD2B) are not available.¹⁰ In this particular group of patients, the DYSF gene - which encodes the dysferlin protein - is deficient or absent, leading to damage to the maintenance of the muscle fiber structure, leading to sarcolemma disruption and motor unit damage, with consequent loss of muscle strength and motor function. Muscle damage during exercise has also been inconclusively associated with changes in muscle excitation-contraction, oxidative, and/or energy metabolism.²¹

Exercise tolerance may be limited due to the above-mentioned pathophysiological characteristics related to the muscle fibers loss or decreased physical activity level associated with motor impairment.²¹ Because of muscular degeneration in dystrophies, tolerance is lower to high-intensity physical exercises and eccentric contractions, inducing muscle damage and subsequent muscle weakness. On the other hand, physical exercise promotes positive effects on several neuromuscular diseases, with no association with susceptibility to muscle injury, and if practiced at mild to moderate intensity, within the limits of the patient tolerance, may prevent atrophy.^{21,22} The present study protocol was based on mild to moderate intensity water-based exercises and promoted physical and functional improvements to the patient, with no complications, adverse effects, or signs of overload in any of the sessions or post-training.

In this context, aquatic physical therapy may be an effective option for patients with LGMD2B. The water immersion provides more significant support and a controlled environment to treat dystrophic patients supported by water physical properties, like density, drag, buoyancy, hydrostatic pressure, and thermodynamics. Among the principles of hydrodynamics, buoyancy stands out, as it can reduce the overload by up to 85% in immersed joints, facilitating patient management and decreasing joint overload.²³ However, despite the potential benefits and advantages of aquatic rehabilitation, to the best of our knowledge, no scientific studies reporting it as a specific therapeutic option to LGMD2B patient's treatment were found.

The patient described in our study has a moderate impairment⁵, with patterns of muscle impairment typically described for limb-girdle muscular dystrophies. Nevertheless, she presents more severe weakness on the lower limbs' muscles - mainly the proximal muscles of the pelvic girdle, thighs, and ankle flexors - leading to more difficulties in activities in need of these muscles, such as standing up a chair or climbing stairs.¹³ Upper limb muscle strength impairment is not in a typical pattern²², with an asymmetrical strength of the right and left deltoid and brachial biceps muscles, keeping symmetry for the other muscles.

An MRI study indicates the replacement of muscle mass by adipose tissue in 88% of the patients, with greater involvement of the posterior region of the thighs and legs.²⁴ Therefore, training with an emphasis on the mentioned muscles was included in the intervention protocol, with adaptations and adjustments on the movements of the main types of swimming, within the patient's functional adaptations in an aquatic environment, performing the movements outside the technical standards.

Functional improvements were assessed after the intervention for muscle strength, functionality, and quality of life. It is noteworthy that the patient had no previous history of physical exercise, sports, or any physical/physiotherapeutic treatments and a sedentary lifestyle, which generates the greater potential for the benefits from physical exercise, even in a short period of intervention. These results agree with previous studies suggesting that a supervised strength training program, associated with low-impact

aerobic exercises such as swimming and stationary cycling, improves cardiovascular performance, muscle efficacy and decreases fatigue in patients with limb-girdle muscular dystrophy.²² Such studies describe aquatic physical therapy interventions of the same duration and frequency in patients with dystrophies¹⁰ and recommend physical exercise and reduction of physical inactivity as part of the treatment.¹⁴

A recently published study evaluated the sensitivity of instruments and measures to assess changes associated with disease progression.¹⁸ It indicated that the total score of the MFM scale is one of the most sensitive instruments for evaluating and monitoring clinical changes in motor function, demonstrating consistent changes throughout time, especially in LGMD2B patients.

Given the slow progression of LGMD2B, establishing treatment and functional rehabilitation options is essential to maintain functional independence for as long as possible. The lack of clinical trials and high-level evidence studies limit the establishment of intervention protocols or therapeutic modalities with assessed efficacy for treating and rehabilitating patients with LGMD2B, adding value to evidence of successful physical therapy treatments, like the one presented in this case report. The evidence currently available indicates that therapeutic physical exercises should be part of the rehabilitation routine of patients with muscular dystrophies, which was confirmed by the findings described in this study, indicating that aquatic physical therapy is safe and effective in treating a patient with limb-girdle muscular dystrophy type 2B.

Future studies with more participants are required to produce evidence to generalize data about the effectiveness of Aquatic Physical Therapy in the scope of care for patients with limb-girdle muscular dystrophy, type 2B (LGMD2B).

Conclusion

The lack of therapeutic strategies to stop the progression of LGMD2B increases the importance of physical therapy treatments to functional rehabilitation, maintenance of functionality, and physical activity in this population.

Functional improvement presented by the patient described here suggests that functional rehabilitation based on aquatic physical therapy, in mild to moderate intensity, was a safe and effective treatment option to gain muscle strength, range of motion, functional capacity, and quality of life of the motor and functional disorders associated with the case described.

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Authors' contributions

Bento-Torres NVO, Luz LES, and Nascimento RCR participated in the conception of the study, intervention delivery, data analysis, interpretation of results, and writing of the manuscript. Jacob MM contributed to the data analysis and writing of the manuscript. All authors read and approved the final version of the manuscript.

Competing interests

No financial, legal, or political conflicts involving third parties (government, corporations and private foundations, etc.) have been declared for any aspect of the submitted work (including, but not limited to grants and funding, advisory council participation, study design, preparation manuscript, statistical analysis, etc.).

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