

Functional evaluation in Chronic Inflammatory Demyelinating Polyneuropathy: a case report

Avaliação funcional na Polineuropatia Desmielinizante Inflamatória Crônica: um relato de caso

Evaluación funcional en la Polineuropatía Desmielinizante Inflamatoria Crónica: un informe de caso

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Resumo

Introdução. A polineuropatia desmielinizante inflamatória crônica (CIDP) é uma doença rara que afeta os nervos periféricos, atingindo a bainha de mielina, causando alterações na força, equilíbrio, mobilidade funcional e sensibilidade. Não existem artigos que descrevam uma avaliação funcional abrangente para a CIDP. **Objetivo.** Descrever o caso de uma paciente diagnosticada com CIDP e fornecer um guia para avaliação funcional. **Relato de Caso.** Uma mulher de 25 anos diagnosticada com CIDP foi avaliada e monitorada por profissionais de fisioterapia em um projeto de extensão universitária. Como manifestação funcional inicial, apresentou dificuldade em usar sapatos abertos, ocorrência de câibras, parestesia bilateral no antepé, desequilíbrio na postura bípede e fraqueza muscular. Para a avaliação funcional, foi proposta a dinamometria manual, o Teste de Pegada de Nove Furos, o sinal de Froment, o teste de levantar e andar, o teste de Romberg clássico e Tandem, a avaliação da sensibilidade tátil, térmica, dolorosa e vibratória e o questionário de qualidade de vida *Short form 36*. Após a avaliação, foram identificados valores abaixo do normal para a força de preensão em ambas as mãos, teste de Romberg positivo, hipoestesia tátil, térmica e vibratória, bem como prejuízo na qualidade de vida. **Conclusão.** A avaliação funcional proposta foi abrangente para contemplar as principais deficiências funcionais e limitações descritas em um caso típico de CIDP, possibilitando o planejamento e a definição de objetivos para a intervenção fisioterapêutica.

Unitermos. Polineuropatia; polineuropatia desmielinizante inflamatória crônica; avaliação fisioterapêutica; capacidade funcional; fisioterapia

Abstract

Introduction. Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare disorder that affects peripheral nerves, targeting myelin sheath, causing changes in strength, balance, functional mobility and sensitivity. No articles describe a comprehensive functional evaluation for CIDP. **Objective.** To describe the case of a patient diagnosed with CIDP and provide a guide for functional assessment. **Case Report.** A 25-year-old woman diagnosed with CIDP was evaluated and monitored by physiotherapy professionals in a university extension project. As an initial functional manifestation, presented impairment for wearing open shoes, cramps occurrence, bilateral paresthesia in the forefoot, imbalance in bipedal stance and muscle weakness. For functional evaluation evaluation was proposed manual dynamometry, Nine-Hole Peg Test, Froment's sign, timed up and go test, classic and Tandem Romberg test, evaluation of tactile, thermal, painful and vibratory sensitivity and Short form 36 quality of life questionnaire. After evaluation, values below normal for grip strength in both hands, positive Romberg test, tactile, thermal, and vibratory hypoesthesia, as well as impairment in quality of life were identified. **Conclusion.** The functional evaluation proposed was comprehensive to contemplate the main functional deficiencies and limitations described in a typical case of CIDP, enabling the planning and definition of objectives for the physical therapeutic intervention.

Keywords. Polyneuropathy; chronic inflammatory demyelinating polyneuropathy; physiotherapeutic evaluation; functional capacity; physical therapy

Resumen

Introducción. La polineuropatía desmielinizante inflamatoria crónica (CIDP) es un trastorno raro que afecta a los nervios periféricos, dirigiéndose a la vaina de mielina y causando cambios en la fuerza, equilibrio, movilidad funcional y sensibilidad. No hay artículos que describan una evaluación funcional integral para la CIDP. **Objetivo.** Describir el caso de una paciente diagnosticada con CIDP y proporcionar una guía para la evaluación funcional.

Informe de caso. Una mujer de 25 años diagnosticada con CIDP fue evaluada y monitoreada por profesionales de fisioterapia en un proyecto de extensión universitaria. Como manifestación funcional inicial, presentó dificultades para usar zapatos abiertos, aparición de calambres, parestesia bilateral en el antepié, desequilibrio en la postura bípeda y debilidad muscular. Para la evaluación funcional se propuso la dinamometría manual, el Test de Pegado de Nueve Agujeros, el signo de Froment, el test de levantarse y andar, el test de Romberg clásico y Tandem, la evaluación de la sensibilidad táctil, térmica, dolorosa y vibratoria y el cuestionario de calidad de vida Short form 36. Después de la evaluación, se identificaron valores por debajo de lo normal para la fuerza de agarre en ambas manos, prueba de Romberg positiva, hipoestesia táctil, térmica y vibratoria, así como deterioro en la calidad de vida. **Conclusión.** La evaluación funcional propuesta fue integral para contemplar las principales deficiencias funcionales y limitaciones descritas en un caso típico de CIDP, permitiendo la planificación y definición de objetivos para la intervención fisioterapéutica.

Palabras clave. Polineuropatía; polineuropatía desmielinizante inflamatoria crónica; evaluación fisioterapéutica; capacidad funcionales; fisioterapia

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INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare autoimmune disease that affects the peripheral nervous system, with an international prevalence

of 0.8–8.9 cases per 100,000 inhabitants¹, affecting all ages and in both sexes, with a prevalence in male adults aged 40 to 60 years^{2,3}.

The CIDP targets the myelin sheath. Its pathogenesis begins when the non-specific antigens of myelinated neurons lead to the production of pro-inflammatory cytokines and interleukins, which permeate the hematoencephalic barrier, allowing the passage of antibodies to the connective tissue that lines the nerve fibers, the endoneurium, causing damage to the myelin and the interruption of nerve conduction².

This polyneuropathy is classified as typical, in which demyelination mainly affects distal nerves and nerve roots, or atypical when demyelination is multifocal in the nerve trunks⁴. CIDP manifestations include several functional changes, such as symmetrical muscle weakness, sensory loss, imbalance, areflexia, fatigue, among others³⁻⁵.

No articles describe a comprehensive functional evaluation for CIDP and studies with physiotherapy intervention are scarce. Among the studies that shine a light on some functional evaluation, Fatigue, Anxiety and Depression^{5,6}, neuromuscular function⁷, deep sensibility; balance; finger dexterity and functional mobility are described³.

Therefore, specific clinical practice studies guiding physiotherapeutic evaluation are relevant and contribute to specialized treatment for functional impairments resulting from the disease. The present study aimed to describe the

case of a patient diagnosed with CIDP and provide a guide for functional assessment.

CASE REPORT

A 25-year-old patient, born in Belém, in the state of Pará, Brazil, diagnosed with chronic inflammatory demyelinating polyneuropathy (CIDP) was evaluated and admitted in February 2020 for physiotherapeutic care in an extension project of Universidade Federal do Pará (UFPA). The project respected the ethical principles for research involving human beings, as stated in resolution 466/12 of the National Health Council, being approved by the Research Ethics Committee of the UFPA under the statement #1,593,170; additionally, the patient signed the Free and Informed Consent Form (ICF) to start treatment.

During the anamnesis, the patient reported that in August 2019 she sought medical attention with complaints of paresthesia and cramps in the forefoot region and imbalance. To confirm the diagnostic hypothesis of CIDP, the patient underwent electroneuromyography, which found an increase in latency in the waves of the F lines of the median and ulnar nerves, and a reduction in the amplitude of the sensory action potentials of the nerves of the upper limbs (UL) and lower limbs (LL). After the diagnosis, she received a drug treatment, including daily intravenous administration of Solu-medrol® (methylprednisolone sodium succinate) for five days and twice-daily ingestion of Etna® capsules (containing 1.5mg uridine, 2.5mg cytidine,

and 1.0mg hydroxocobalamin) for one month to aid in peripheral nerve repair.

The evaluation also considered the International Classification of Functioning, Disability and Health (ICF) to characterize which structures and functions were affected, which activities were compromised by the patient's condition, and what were the effects on her participation. As a result, during the physiotherapy evaluation, it was found that the patient reported difficulty wearing open shoes (d6180), experiencing issues such as splitting, limited to using shoes with heel and ankle support. She also described paresthesia (b265), cramps in the forefoot region, increasing over time, imbalance (b2351), and weakness in lower limbs (b7303) for activities like climbing stairs (d4550) and transitioning from sitting to standing (d4570). Additionally, there's an impact on her daily routine (d230), raising concerns about potential effects on her quality of life.

RESULTS

A functional evaluation was constructed based on clinical rationale, considering complaints, signs, symptoms, pathophysiology, and the biopsychosocial model. Specific tests included the Medical Research Council (MRC) for general muscle strength, manual dynamometry for handgrip strength, Nine-Hole Peg Test (9HPT) for manual dexterity, Froment, Phalen, and Tinel signs for nerve integrity, clinical tests for superficial and deep sensitivity,

Timed Up and Go test (TUG) for lower limb mobility, traditional and Tandem Romberg tests for static balance, and QoL evaluation using the SF-36 questionnaire.

Muscle strength in upper and lower limbs was assessed, with any muscle graded below 4. Results are presented in Table 1. Handgrip strength was measured using a manual dynamometer (Saehan - SH5001). Lower scores are linked to falls, disability, fatigue, functional decline, and impaired health-related quality of life^{8,9}. The test started on the dominant side with the patient sitting on a standardized chair, shoulders adducted, elbows flexed at 90°, and forearms and wrists in a neutral position. The patient was instructed to grip the device with maximum force. Three attempts were made for each side, with a 1-minute interval between attempts. The result was equivalent to the higher value of each side⁹.

Due to complaints of the right UL reported by the patient, manual dexterity was evaluated by the 9HPT. The patient inserted and removed nine pins as quickly as possible using one hand, while the other hand remained static on the device. Time for each limb was assessed, and the result was based on the average time from two attempts for both pin placement and removal¹⁰.

The ulnar nerve function, including pinch strength, was assessed using the Froment Sign. The patient held a sheet of paper between her thumb and index fingers while the examiner attempted to pull it, revealing nerve analysis. A positive result indicated compensation during paper

removal, indicating weakness in the adductor pollicis muscle¹¹. The results obtained from the evaluation of the upper limbs are shown in Table 2.

Table 1. Results of the MRC evaluation for general muscle strength

Joint	Movements	Right Side	Left Side
Shoulder	Flexion	4	4
	Extension	4	4
	Abduction	5	5
	Adduction	4	4
	Internal Rotation	5	5
	External Rotation	5	5
Elbow	Flexion	5	5
	Extension	5	4
Fist	Flexion	5	5
	Extension	5	5
Hip	Flexion	4	4
	Extension	4	4
	Abduction	4	4
	Adduction	4	4
	Internal Rotation	5	5
	External Rotation	4	4
Knee	Flexion	4	4
	Extension	5	5
Ankle	Dorsiflexion	4	4
	Plantarflexion	5	4

Table 2. Results of the evaluation of upper limbs. Values were obtained by dynamometry, Nine-Hole Peg Test and Froment signal tests.

clinical trials	evaluation results	normality
Dynamometry		
Right hand	18.14 kgf	25.3±0.28 kgf
Left hand	17.23 kgf	24±0.26 kgf
9HPT		
Right hand	19.13 sec	15.90±1.91sec
Left hand	25.21 sec	16.97±1.77 sec
Froment's sign		
Right hand	Positive	Negative
Left hand	Negative	Negative

Normality values were obtained for dynamometry⁹ and the nine-hole peg test¹⁷.

The functional mobility of the lower limbs was assessed by the TUG. This test was performed due to complaints of mobility and weakness in the LL presented by the patient, representing a risk of fall. The test involved getting up from an armchair, walking to a cone three meters away, turning around, and returning to sit. The evaluator conducted the test, with the first attempt serving as a practice to ensure familiarity. Slower TUG scores are associated with worse perceived physical and mental health¹². The functional mobility results are described in Table 3.

The Romberg test was performed to evaluate static balance. This test was chosen due to the imbalance, weakness and paresthesia in the forefoot reported by the patient. The classic test was done with feet together, first with eyes open for 60 seconds and then with eyes closed for the same duration. Following that, the Tandem Romberg test was conducted, initially with eyes open for 60 seconds, and then with eyes closed for the same period. In both tests, any attempt by the patient to reposition the foot, seek support, or open the eyes in the closed eyes test would be considered a positive result, in line with¹³. The results of the functional mobility and balance tests are shown in Table 3.

Superficial and deep sensitivity were assessed due to reduced sensory action potential amplitudes in both upper and lower limbs. Tactile, painful, and thermal sensitivity were evaluated using cotton, a wooden toothpick, and test

tubes containing cold and warm water, this last assessment showed a low sensitivity in the S1 region. The evaluation involved stimulating the patient's upper and lower limb dermatomes using these tools. Vibratory sensitivity was also assessed with a tuning fork on the hallux bilaterally, indicating reduced sensitivity in the lower limbs and a lack of sensitivity in the S1 region. Finally, the Phalen and Tinel signs were conducted to rule out compressive neuropathy. Phalen's sign involved flexing the wrists for 60 seconds, and Tinel's sign was performed by tapping over the median nerve at the wrist. Both tests are positive if numbness, paresthesia, or tingling occur in the median nerve distribution¹⁴. The results of the evaluation of each type of sensitivity are described in Table 3.

Table 3. Evaluation of functional mobility and balance evaluated with the Timed up and Go (TUG), Romberg and Tandem Romberg tests superficial (tactile, thermal and painful) and deep (vibratory) sensitivities, Phalen and Tinel’s signs.

clinical tests	results	normality
TUG	9.5 sec	8.57±1.40 sec
ROMBERG	Positive	Negative
TANDEM ROMBERG	Positive	Negative
TYPES OF SENSITIVITY	results	
TACTILE SENSITIVITY	Normoesthesia	
THERMAL SENSITIVITY	Hypoesthesia (L5-S1)	
PAINFUL SENSITIVITY	Normoesthesia	
VIBRATORY SENSITIVITY	Hypoesthesia (S1)	
PHALEN'S SIGN	Negative	
TINEL 'S SIGN	Negative	

Normality values obtained for TUG¹².

For QoL assessment, the patient filled out the Brazilian version of the Medical Outcomes Study 36 - Item Short Form Health Survey (SF-36) questionnaire. Given the symptoms and functional impairments, the patient might have a low quality of life, justifying the use of this instrument. The questionnaire comprises 36 questions across eight domains (functional capacity, physical aspects, pain, general health status, vitality, social aspects, emotional aspects, and mental health). Each domain was analyzed separately, with scores ranging from zero to one hundred points, where higher scores indicate better QoL^{15,16}. The score suggests which aspects of the patient's life are affected by the CIDP and the severity. The results obtained with the SF-36 questionnaire are shown in Table 4.

Table 4. Quality of life assessed with the SF-36 questionnaire.

SF-36 DOMAINS	SCORE
FUNCTIONAL CAPACITY	25
PHYSICAL ASPECTS	50
PAIN	62
GENERAL HEALTH STATUS	27
VITALITY	60
SOCIAL ASPECTS	25
EMOTIONAL ASPECTS	33.33
MENTAL HEALTH	64

DISCUSSION

In this report, we described a case of a young female with typical CIDP and the purpose of physiotherapeutic evaluation to find functional deficits. The frequency of CIDP is higher in male individuals over 40 years of age², a different profile from the case reported.

The most frequent symptoms of CIDP described in the literature correspond to proximal and distal muscle weakness, predominantly distal, decreased sensory function, areflexia, decreased balance, impaired gait phases and fatigue³.

Regarding proximal muscle strength, the normal values for handgrip in healthy women aged 20 years or older were 25.3 ± 0.28 kgf and 24 ± 0.26 kgf for the right and left hands, respectively⁹. Thus, the patient in this report has bilateral muscle weakness, with a reduction of approximately 30% in strength. This seems to be indicative that physical therapy should consider exercises for strength and resistance^{5,16}.

The patient showed a reduction in manual dexterity, in both hands, especially in the left hand, since the 9HPT was performed at a time of 19.13s and 25.1s by the right and left hands, respectively. The expected mean normality values for healthy women, of the same age group, are 15.90 ± 1.91 s for the right and 16.97 ± 1.77 s for the left hand¹⁷. Low manual dexterity scores in the 9HPT are related to the impact on daily living activities, work, and self-care activities¹⁰.

In the test of the integrity of the ulnar nerve by Froment's sign, the patient presented compensation of movement only in the right hand, revealing the origin of the limitation on the right hand to hold objects. The insufficiency of the adductor pollicis muscle impairs the action of "holding the role" during its removal by the evaluator, being compensated by the activation of the long flexor of the thumb, innervated by the median nerve¹³.

Regarding the integrity of nerves, hypoesthesia is usually a frequent feature in patients with CIDP and this is in accordance with our sensorial evaluation^{3,5}. Although four sensory modalities were evaluated, only pain was preserved. There are no specific articles about sensorial modalities impaired and sensorial rehabilitation in patients with CIDP, but sensorial rehabilitation seems effective in functionality for other neural injuries¹⁸.

For the static balance analysis, the patient underwent the classic Romberg and Sharpened Romberg tests presented an increase of postural instability when eyes are closed, and this occurs in patients with severely compromised deep sensitivity¹⁹. Currently, no studies have examined the test-retest reliability of the Romberg test for subjects with CIDP but balance and proprioception training should be included in the physiotherapy program.

Regarding functional mobility by TUG, the patient performed the test in agreement with the normative reference values¹², which demonstrated that a time of less than 10 sec is expected for healthy people.

The result of the SF-36 questionnaire showed that the patient had a QoL score considered low for all domains if compared to women between 25 and 34 years of age¹⁵. Such differences are even more discrepant when analyzing the domains of functional capacity and social aspects since the results found in this report are lower than 70%. Patients with stable CIDP have significantly lower SF-36 scores if compared to healthy individuals, suggesting that many capacities and, performance are compromised²⁰.

The present study shows some limitations since fatigue, one of the clinical manifestations often found in patients with PDIC³, was not directly measured. In another way, the domain "Vitality" of the SF-36 shows issues related to fatigue and helps to understand this manifestation. Thus, the inclusion of instruments such as the fatigue severity scale is recommended for a complete evaluation.

CONCLUSION

The applied functional assessment guide was effective in evaluating the most frequent functional limitations described in a typical case of PDIC (which stands for something not specified), as it allowed the observation of bilateral muscle weakness, limited manual dexterity, sensory alteration, and reduced quality of life in the assessed patient. It was deemed necessary to include a more specific tool for evaluating fatigue. Therefore, the

applied functional assessment guide enables the planning and setting of objectives for physiotherapeutic intervention.

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