

Chronic inflammatory demyelinating polyneuropathy: quality of life, sociodemographic profile and physical complaints

Polineuropatia inflamatória desmielinizante crônica: qualidade de vida, perfil sociodemográfico e queixas físicas

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ABSTRACT

Whereas an evaluation of quality of life and possible impacts on the mental state of a patient may help to evaluate the evolution of chronic inflammatory demyelinating polyneuropathy (CIDP), the aim of this study was to study the psychological profile of patients, and evaluate quality of life associated with the disease. **Method:** 41 patients were evaluated using a Mini-Mental State Examination (MMSE) and a Short-Form Health Survey (SF-36). **Results:** The mean age of the patients was 50.6 years, 63.4% men. Of the participants, 65.9% had other health problems, 39% reported needing help with activities of daily living, 49% slept less than 8 hours per night, and 34.1% complained of some memory deficit. The average MMSE score was 26. Impairment of functional capacity and pain were the more important altered health states. **Conclusion:** CIDP has important social and economic impacts, owing to functional impairments that can lead to professional and personal limitations.

Keywords: chronic inflammatory demyelinating polyradiculoneuropathy, CIDP, quality of life.

RESUMO

A avaliação da qualidade de vida (QV) e dos possíveis impactos dos déficits funcionais sobre o estado mental de pacientes com polirradiculoneuropatia inflamatória desmielinizante crônica (PIDC) pode contribuir para a melhor compreensão de aspectos evolutivos da doença. A presente investigação teve como objetivo estudar as atividades da vida diária de pacientes com PIDC e avaliar a sua QV. **Método:** Foram avaliados 41 pacientes através do Mini Exame do Estado Mental (MEEM) e do inventário de saúde SF-36*. **Resultados:** A média de idade dos participantes foi 50,6 anos, 63,4% homens. Problemas adicionais de saúde foram referidos por 65,9%: 39% relataram necessitar de ajuda para atividades de vida diária, 49% dormiam menos de 8 horas por noite e 34,1% referiam alguma dificuldade de memória. A média do MEEM foi 26. Através do SF-36 foi verificado maior prejuízo na capacidade funcional; a referência a dor foi proeminente. **Conclusão:** A PIDC pode ter importante impacto social e econômico em decorrência dos prejuízos funcionais primários e secundários que podem levar ao afastamento do trabalho.

Palavras-chave: polirradiculoneuropatia, CIDP, qualidade de vida.

Chronic inflammatory demyelinating polyneuropathy (CIDP) is considered to be an autoimmune disease of the peripheral nervous system, characterized by symmetrical weakness of the distal and proximal muscles, which increases progressively and persists for more than two

months. It is associated with reduced sensitivity, lack or reduction of tendon reflexes, elevated protein levels in the cerebrospinal fluid, changes in nerve conduction due to demyelination, and signs of demyelination also in a nerve biopsy. CIDP may present in the recurrent

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form (with relapses) or in the chronic and progressive form^{1,2}.

Axonal loss may occur as a result of demyelination, leading to functional damage and a variable extent of physical symptoms that may impair the performance of daily life activities. Thus, evaluation of the magnitude of axonal loss is relevant to the prognosis of the disease, and the assessment of a positive response to treatment should consider improvement in patient strength, sensitivity and functional capacity².

Since CIDP is a chronic condition that may have a progressive course leading to physical and functional limitations, an evaluation of quality of life and of the possible impact of the disease on the mental state of the patient can contribute to an understanding of the evolution of the disease and its prognosis.

Studies have suggested the importance of evaluating the quality of life of patients in order to complete an assessment of CIDP since this information would expand the doctor's knowledge about the functional limitations of his/her patient and the psychosocial impairment caused by the disease³.

Evaluation of the quality of life of patients with CIDP has shown that this condition leads to greater impairment of functional capacity and physical health conditions than emotional and social aspects³⁻⁶. In addition, it seems that these physical limitations do not interfere with the emotional and social functioning of the patients⁴.

Regarding the instruments for evaluation, a study on patients with CIDP demonstrated that self-report measures about physical functions are as effective as clinical measures, although it was suggested that the most appropriate instruments for the evaluation of body function may vary according to the type of neuropathy studied¹.

In view of the fact that an evaluation of quality of life and of the possible impacts on the mental status of the patient can contribute to an understanding of the evolution of the disease and its prognosis, the objective of the present study was to determine the sociodemographic and health-related quality of life profile of patients with CIDP managed by a general hospital.

METHOD

Study design and participants

This is a descriptive cross-sectional study approved by the Research Ethics Committee of the hospital in agreement with the Declaration of Helsinki regarding research on humans. Initial contact with the patient was made on the day of a routine visit to an outpatient clinic or during hospitalization, together with an evaluation of the patient. Participation in the study was random. All patients gave written informed consent to participate.

In this study, 41 out of 112 patients diagnosed with CIDP (and seen at the same clinic) were evaluated at the Neuro-muscular Diseases Outpatient Clinic of the University Hospital of the Ribeirao Preto School of Medicine, University of Sao Paulo, from October 2009 to February 2012. We excluded three patients younger than 18 years and one patient with another suspected diagnosis that might have excluded CIDP if confirmed. The CIDP diagnosis was made in accordance with the European Federation of Neurological Societies and the Peripheral Nerve Society Guidelines⁷.

The mean age of the participants was 50.6 years (range: 26 to 76 years) and the mean duration of the disease was 5.7 years at the time of evaluation.

The sample consisted of 26 (63.4%) men and 15 (36.6%) women. Thirty-five patients (85.4%) declared that they followed a religion and only 11 (26.8%) were professionally active. Most participants had at least 8 years of schooling (58.5%) and cohabited with a companion (73.2%). Two participants could not respond to the SF-36 questionnaire owing to their time limitation.

Instruments

The patients were interviewed in order to gather sociodemographic data and information about the disease, i.e., information about sex, age, schooling, religion, housing conditions, habits and life routine, health problems, time of onset of symptoms, time since diagnosis, and previous and current treatments.

The Mini Mental State Examination (MMSE) was applied to evaluate cognitive abilities. The MMSE investigates temporal and spatial orientation, memory and language, with a score ranging from 0 to 30. The lower the score, the worse the cognitive status of the patient.

A 36-item Short-Form Health Survey (SF-36) (which originally formed part of the Health Outcomes Study) was applied in order to assess quality of life. This is a multidimensional questionnaire consisting of 36 items divided into eight categories: functional capacity, physical aspect, pain, general health status, vitality, social aspects, emotional aspects, and mental health.

RESULTS

General health

The information about general health is summarized in Table 1. It can be seen that 27 (65.9%) participants had some health problem in addition to CIDP; 16 (39%) reported that they needed help to perform some daily life activities (bathing, getting dressed, going to the bathroom, transferring from the bed or the chair, controlling waste elimination, eating); 20 (49%) slept less than 8 hours per night, and 14

Table 1. General health and daily activities.

Patient information	No. of cases (%)
Health problems other than CIDP	27 (65.9)
Needs help with ADL*	16 (39.0)
Night sleep:	
Sleeps 2–4 hours per night	7 (17.5)
Sleeps 5–7 hours per night	13 (31.7)
Sleeps 8–12 hours per night	21 (51.2)
Daytime sleep	14 (34.1)
Memory deficit	14 (34.1)
Smokes	11 (26.8)
Drinks alcoholic beverages	13 (31.7)
Routine:	
Active	33 (80.5)
Inactive**	8 (19.5)
Physical activity or physiotherapy	21 (51.2)
Leisure activities	26 (63.4)

*ADL: activities of daily life; **Does not perform any physical activity

(34.1%) reported some memory difficulties. In addition, habits such as smoking and drinking alcohol were present, as well as physical and leisure activities.

Regarding the presence of other health problems, 12 (29.3%) participants had arterial hypertension, 12 (29.3%) had diabetes mellitus, 4 (9.8%) had hypothyroidism, and 14 (34.1%) had other diseases (each reported a different disease, and one stated that he had motor neuron disease).

Chronic inflammatory demyelinating polyneuropathy

The mean duration of CIDP (since the onset of symptoms) was 7.25 years (2-12) and the mean time since diagnosis was 5.74 years (1-10); 15 (36.6%) patients were under pharmacological treatment at the time of evaluation. The disease started exclusively with symptoms in the lower limbs (feet or legs) in 18 (43.9%) patients, in the lower and upper limbs in 17 (41.5%), and exclusively in the upper limbs (hands and arms) in 2 (4.9%), with 4 (9.8%) being unable to provide this information. More than half the participants (58.5%) reported that they had undergone some other type of treatment before receiving a diagnosis of CIDP, and 5 (12.2%) associated the onset of symptoms with a stressful event in their life. Table 2 presents the patients' complaints at the time of evaluation.

Table 2. Complaints at the time of evaluation.

Complaint	No. of cases (%)
Weakness, fatigue or tiredness	14 (34.1%)
Paresthesias	12 (29.3%)
Pain	20 (48.8%)
Difficulty in walking or moving around	16 (39%)
Tripping or falling	2 (4.9%)
Lack of sensitivity or numbness	6 (14.6%)
Cramps	8 (19.5%)
Other	4 (9.8%)

Mental state and quality of life

The mean MMSE score was 26 (22-30) and the results of the SF-36 indicated greater impairment of functional capacity (mean 40.64) and pain (mean 58.68) (Table 3).

DISCUSSION

The patients evaluated represented a little more than 40% of all patients with a CIDP diagnosis seen in a neuromuscular outpatient clinic of a CIDP referral center. Considering the low incidence of CIDP in the population, and despite the small number of participants, it was possible to outline the profile of these patients, favoring more information about the CIDP patients and raising new possibilities of investigation of the disease.

As also reported in studies conducted in other countries, CIDP patients are aged about 50 years old and the prevalence is twice as high among men^{4,8,9}. So far, there are no scientific data to explain this male predominance, nor any hypothesis.

The mean age of the participants in our study was also similar to that reported in other studies, showing that CIDP affects a middle-aged population, with important socioeconomic impacts since the disease can impair the functional capacity of the patients, consequently leading to work losses.

The rate of professionally active patients (26.8%) demonstrates the socioeconomic impact of CIDP, despite a low overall incidence. Prevalence rates of 0.15-5 per 100,000 individuals have been found in developed countries¹⁰. There is an obvious importance to expanding our knowledge about the clinical course of the disease and the associated functional and quality of life changes.

There appear to be no previous studies concerning the level of activity of patients with CIDP. Although some studies evaluated functional capacity, they did not mention work activity, leisure or physical activity. One exception was a study that aimed to determine the impact of physical activity on muscle strength¹¹.

If we compare the information about daily physical activity (80.5% of the patients reported an active routine, with activities inside and outside home) with the number of professionally active patients (26.8%) in the present study, we may question whether some of these patients could have returned to the job market or even never have abandoned it if patients with CIDP received a more health-integrated care package (complete diagnostic tests, physiotherapy and physical activity, appropriate pharmacotherapy) and an earlier diagnosis. It is known that, with treatment, 60% of patients have a monophasic or recurrent form of the disease, while 40% have a progressive form¹².

Regarding general health, almost two-thirds of the patients reported some other problems, including arterial hypertension and diabetes mellitus (DM). Although some

Table 3. Quality of life.

SF-36 category (n=39)	Mean (±SD)	Median	Mode
Functional capacity	40.64 (26.41)	40	40
Physical aspects	65.38 (38.74)	75	100
Pain	58.68 (28.68)	52	100
General health status	60.26 (23.8)	57	72
Vitality	61.28 (21.14)	65	40
Social aspects	61.91 (31.24)	62.5	100
Emotional aspects	68.97 (37.89)	100	100
Mental health	67.08 (23.35)	76	76

studies have pointed out the co-occurrence of CIDP and DM, we found no evidence of an association between the two diseases. In our sample, we detected a rate of DM incidence that was almost five times higher than that reported in an epidemiological study conducted in a North American city¹³, namely 29.3% compared with 4% in the cited study.

Regarding the co-occurrence of hypertension, we found no information in the literature, except that hypertension has been reported to be a side-effect of treatment with intravenous immunoglobulin¹⁴.

With respect to the use of tobacco and alcohol, the rate of smoking observed in the present study was higher than the prevalence rate for Brazil as a whole (17.2%), a fact that may be possibly explained by the larger number of men in our sample, since cigarette smoking is higher among men than among women¹⁵.

Specifically regarding CIDP, we observed an interval of one and a half years between the mean time of disease onset (the start of symptoms reported by the patient) and the mean time of diagnosis. This difference may result from a variety of factors: a delay in the patient seeking health advice or a delay in being referred for specialized evaluation; scarcity or absence of equipment and of qualified personnel in the services to which the patient was referred for specialized exams, e.g., electroneuromyography or nerve biopsy; difficulty in reaching a differential diagnosis with Guillain-Barré syndrome and Lewis-Sumner syndrome; and a variety of subtypes of the disease, also impairing a differential diagnosis of CIDP^{2,16}.

The abovementioned conditions may explain the large number of patients who stated that they had undergone other treatments before receiving their diagnosis.

Almost all patients reported initial symptoms affecting the lower limbs. Although this is characteristic of the disease, it is possible that impaired walking is more often remembered by the patients because it has a greater impact on their quality of life and on their activities and relationships since it can limit both mobility and independence. After evaluation by the neurology service, almost half of the patients in our study (18 of 41) had difficulty in walking and moving; they also experienced tripping and falls.

The results of the MMSE were similar to those reported in another study on the Brazilian population¹⁷, and the

Brazilian cut-off scores were as expected for the MMSE (13 for illiterates, 18 for low/middle school age and 26 for high school age)¹⁸. However, even though cognitive screening did not suggest impairment in cognitive function, one-third of the patients reported memory deficits, a fact that needs further investigation, with special emphasis on this function.

Regarding quality of life, the results for the patients in our study were similar to those obtained in other studies, though slightly better for hypertensive patients in the following categories: physical aspects, pain, general health status, emotional aspects and mental health¹⁸. The greatest difference in results was observed in the functional capacity category – 40.64 for CIDP patients and 66.28 for hypertensive patients. Functional capacity data were quite similar to those obtained for patients with rheumatoid arthritis¹⁹. It is possible that our CIDP patients had specialized neurological evaluation or received a diagnosis only when functional impairment was already significant. It should be borne in mind that perhaps most of the patients evaluated had progressive CIDP with consequent lower results regarding functional capacity. It should be remembered that in the present study we did not determine whether CIDP was monophasic, recurrent or progressive.

Compared with one Danish study⁴, our patients showed greater impairment of quality of life, except in the categories of general health status and vitality. However, the Danish study evaluated 14 patients, whereas our study evaluated 41. Furthermore, the differences may also be related to cultural differences since the quality of life construct is not objective and the data were derived from a self-report measure.

Regarding health questions, many patients reported sleep and memory difficulties. We may hypothesize that paresthesias and pain interfere with the sleep of the patients. However, a more in-depth evaluation is necessary, also to determine the need for a direct intervention regarding this problem. Improved sleep may favor quality of life for patients with CIDP. Similarly, new investigations may clarify whether or not there is a link between the memory difficulties reported by patients and the disease or its treatment. It is possible that, facing such uncomfortable symptoms as paresthesias, weakness or difficulty in walking or moving the hands, the patients dismiss small memory difficulties or even other non-physical symptoms. We have not been able to find any reports in the literature about the association between memory and CIDP, or about a possible impact of this condition on sleep.

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