Efficacy of long-term prednisone therapy in patients with chronic inflammatory demyelinating polyneuropathy (CIDP): a retrospective cohort study

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Introduction. Patients with CIDP respond adequately to steroid therapy and intravenous immunoglobulin (IVIG). However, few patients have access to IVIG in developing countries. Little information exists about the clinical response to steroid therapy in Latin American countries.

Objective. to describe the long-term functional clinical response (24 months) to prednisone therapy in CIDP patients.

Material and methods. A retrospective cohort was conducted. Selection included patients with definitive CIDP diagnosis according to European criteria from the Neuromuscular Diseases clinic of the National Institute of Neurology and Neurosurgery between January 2016 and December 2020. Good response to steroid therapy was defined as with improvement in at least one point on the GBS disability score. Poor response to steroid therapy was defined as patients who did not show improvement in at least one point on the GBS disability score. Patients were evaluated at 3, 6, 12, 18 and 24 months.

Results. Forty-seven patients with CIDP were included. Half of them were male and mean age was 46±15 years. Mean time since symptom onset to diagnosis was 6 (IQR 2-12) months. The most common clinical variant was sensory-motor 57.4%, followed by acute-onset CIDP 21.3% and atypical variants 21.2%. At diagnosis our patients presented: mean GBS disability score of 3 (2.25-4) points, MRC score 39.5 ± 12 points, independent gait in 17%, mean prednisone dose of 50 mg (32.5-50). Twenty-four months after prednisone therapy, a less mean GBS disability score –1(0-2) points–, mean MRC score 56.3 ± 5.1 points, independent gait 93% and prednisone dose 1 (0-5) mg. Patients with poor three-month functional clinical response had a delay in diagnosis > 6 months (64.7% vs 27.5%) and atypical clinical variants (47% vs 6.8%).

Conclusion. CIDP patients treated with prednisone have good long-term functional clinical response. Delay in diagnosis and atypical variant are common clinical characteristics for poor functional clinical response in treatment with prednisone.

Key words. Chronic. CIDP. Neuropathy. Prednisone. Prognosis. Treatment.

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Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is the most common acquired chronic inflammatory neuropathy, with an incidence of 0.15-10.5 cases per 100,000 inhabitants/year and with a male predominance [1]. Typical CIDP accounts for 50-60% of CIDP cases and manifests as a progressive (> 8 weeks) neuropathy with proximal and distal limb weakness, sensory disturbances and decreased or absent muscle stretch reflexes. Contrary to Guillain-Barré syndrome, typical CIDP rarely affects cranial nerves [2]. Atypical variants include asymmetric, sensory predominant, distal predominant and motor predominant [3].

The European Federation of Neurological Societies and the Peripheral Nerve Society criteria are the most widely used for CIDP diagnosis, due to its high sensitivity and specificity. These are based on the combination of clinical and electrophysiological findings [4]. Recent research also supports magnetic resonance imaging and peripheral nerve ultrasound as ancillary tests [5].

Patients with CIDP respond adequately to steroids and intravenous immunoglobulin (IVIg) in 85% of cases, while 15% are treatment refractory [6]. Monthly IVIg is expensive in developing countries, few people have access to this type of treatment. Because of this, steroid therapy has demonstrated to be effective for CIDP, although adverse effects are fre-

Table I. Baseline patients'characteristics.

	n = 47
Age (years), median (SD)	46 ± 15
Male, *n* (%)	26 (55.3)
Comorbidities:	
Hypertension, *n* (%)	5 (10.6)
Diabetes, *n* (%).	9 (19.1)
Autoimmune desease, *n* (%)	2 (4.3)
Time from symptom onset to diagnosis (months), median (IQR)	6 (2-12)
MRC score at diagnosis, median (DS)	29.3 ± 21.9
GBS disability scale, median (IQR)	3 (3-4)
Cranial nerve involvement, *n* (%)	6 (12.8)
Facial, *n* (%)	4 (8.5)
Bulbar, *n* (%)	4 (8.5)
CIDP typical variants:	
Sensory-motor, *n* (%)	27 (57.4)
Acute-onset CIDP, *n* (%)	10 (21.3)
Atypical variants:	
Distal predominant, *n* (%)	4 (8.5)
Assymetrical, *n* (%)	2 (4.2)
Ataxia-tremor, *n* (%)	2 (4.2)
Sensory predominant (%)	2 (4.2)
Albuminocytological dissociation, *n* (%)	40/44 (90.9)
Proteins (mg/dL), median (IQR)	124 (60-245)
Treatment at diagnosis:	
Prednisone, *n* (%)	14 (29.8)
Prednisone + methylprednisolone pulses, *n* (%)	22 (46.9)
Prednisone + plasma exchange or IVIg, *n* (%)	10 (21.3)
Steroid-sparing agent (Azatioprina, ciclofosfamida), *n* (%)	22 (46.8)
Time to initiation of steroid-sparing agent, months (RIQ)	9 (1-18)

CIDP: chronic inflammatory demyelinating polyneuropathy; GBS: Guillain-Barre Disability Score; IQR: interquartile range; IVIg: intravenous immunoglobulin; MRC: Medical Research Council; SD: standard deviation.

quently reported [7]. The aim of the study is to describe the CIDP population of a Latin American center, and the long-term effects of steroid therapy.

Material and methods

A retrospective cohort was conducted. Selection included patients with definitive CIDP diagnosis according to European criteria [4] from the Neuromuscular Diseases Clinic of the National Institute of Neurology and Neurosurgery between January 2016 and December 2020. Additionally, all patients with prednisone treatment at diagnosis and during follow-up were included. We obtained the following data: age, sex, comorbidities, Guillain-Barre Disability Score [8], Medical Research Council Score, time since symptom onset to diagnosis (months), cranial nerve involvement, albuminocytological dissociation and CSF proteins. Patients were clinically classified as: typical (sensory-motor or subacute onset), atypical phenotype (asymmetric, distal predominance, motor predominance, sensory predominance or ataxia/tremor).

The standard initial prednisone dose was 50 mg in most patients, while dose reduction was adjusted to 5-10 mg depending on clinical response monthly. We also registered if the patient received other treatments (methylprednisolone pulses, IVIg, plasma exchange) apart from prednisone therapy or steroid-sparing agents such as azathioprine, cyclophosphamide, methotrexate and mycophenolate. In the follow-up time evaluation at 3, 6, 12, 18 and 24 months, the following data was obtained: prednisone dose, MRC score, independent gait and GBS disability score. In addition, presence of relapse was defined as increase in at least 1 point on the GBS disability score. Good response to steroid at three months of follow-up was defined as patients with improvement in at least one point on the GBS disability score. Poor response to steroid therapy was defined as patients with no improvement in at least one point on the GBS disability score.

Statistical analysis

For the descriptive analysis, the distribution of continuous variables was determined with the Kolmogorov-Smirnov test. Variables were described as means, standard deviation (SD) or medians and interquartile range according to their distribution. The categorical variables were described in frequencies and percentages. To examine differences between groups, the following were used: χ^2 test

and Fisher's exact test for categorical variables, Student's *t* test or ANOVA to compare means, and Mann-Whitney or Kruskall Wallis *U* test to compare medians. Value of $p < 0.05$ was considered statistically significant. All statistical analyzes were done with SPSS version 22.0.

Results

Forty-seven patients were included in this study. Fifty-five percent were male, and the median age was 46 ± 15 years. The most frequent comorbidity was diabetes (19.1%). Time since symptom onset to diagnosis (median) was 6 (IQR 2-12) months, while the MRC score at diagnosis was 29.3 ± 21.9 points. GBS disability score at diagnosis (median) was 3 (IQR3-4) points. Thirteen percent had cranial nerve involvement and 17% were able to walk independently at diagnosis.

The most frequent clinical variant was the classic sensory-motor variant (57.4%), followed by acute-onset CIDP (21.3%). Regarding CSF findings, 90.9% had albuminocytologic dissociation at diagnosis, with CSF proteins of 124 (IQR 60-245) mg/dL. At diagnosis, 29.8% were treated with prednisone, 46.9% with prednisone and methylprednisolone pulses and 21.3% with plasma exchange or IVIg preceding prednisone. Clinical features are summarized in Table I.

Atypical variants were reported in 21.2%, with a significant delay in diagnosis compared with typical variants, 5 (IQR 2-18) vs 10.5 (IQR 8-24) months $p = 0.002$. In addition, 80% with atypical variants presented albuminocytologic dissociation, with less CSF protein levels compared with typical variants –152 (IQR 50-250) vs 106 (IQR 67.7-177) mg/dL– $p = 0.819$.

In the analysis comparing patients with good response vs patients with poor response at 3 months, we found significant differences delay in diagnosis > 6 months (64.7% vs 27.5%) and atypical clinical variant (47% vs 6.8%) (Table II).

Thirty-two patients were followed for 24 months. Fifty-eight percent had at least one symptom relapse at 18 (IQR 3-18) months. In the follow up at 24 months, we found a good clinical response as prednisone dose was reduced, as well as improvement in GBS disability score (Fig. 1), MRC score (Fig. 2) and in independent gait (Table III).

Twenty-two patients had combined therapy with steroid-sparing agents (21 with AZT 1-2mg/kg and one patient with monthly cyclophosphamide. Forty percent presented adverse effects. Twenty-three reported weight gain and obesity, 12.7% Cushing syndrome, 14.5% hyperglucemia, 6.3% anxiety, 4.2% gastritis and 2.3% glaucoma.

Table II. Clinical differences in patients with good vs poor treatment response at three month follow up since steroid therapy initiation.

	Good response *n* = 29	Poor response *n* = 17	*p* value
Age (años), median (DS)	45.8 ± 16.7	45.7 ± 12.4	0.97
Age > 60 years, *n* (%)	6 (20.6)	(17.6)	> 0.99
Male, *n* (%)	18 (62)	8 (47)	0.36
Diabetes, *n* (%).	4 (13.7)	4 (23.5)	0.44
Time since symptom onset to diagnosis (months), median (IQR).	4 (1.5-9)	8 (5.5-18)	0.005
Delay ≥ 6 months to diagnosis, *n* (%)	8 (27.5)	11 (64.7)	0.028
MRC score at diagnosis (SD)	37.6 ± 12.9	40.12 ± 11.9	0.52
Cranial nerve involvement *n* (%)	2 (6.8)	3 (17.6)	0.34
Typical variant, *n* (%)	27 (93.1)	9 (52.9)	0.003
Sensory-motor, *n* (%)	18 (62)	8 (47)	0.36
Acute-onset CIDP, *n* (%)	9 (31)	1 (5.8)	0.047
Atypical CIDP variants, *n* (%)	2 (6.8)	8 (47)	0.003
Distal predominant, *n* (%)	1 (3.4)	3 (17.6)	0.53
Assymetrical, *n* (%)	0 (0)	2 (11.7)	
Ataxia-tremor, *n* (%)	1 (3.4)	1 (5.8)	
Sensory predominant *n* (%)	0 (0)	2 (11.7)	
Treatment at diagnosis:			
Prednisone, *n* (%)	10 (34.4)	(29.4)	> 0.99
Prednisone + methylprednisolone pulses, *n* (%)	13 (44.8)	(52.9)	> 0.99
Prednisone + plasma exchange or IVIg, *n* (%)	6 (20.6)	3 (17.6)	0.73

CIDP: chronic inflammatory demyelinating polyneuropathy; GBS: Guillain-Barre Disability Score; IQR: interquartile range; IVIg: intravenous immunoglobulin; MRC: Medical Research Council; SD: standard deviation.

Discussion

Chronic inflammatory demyelinating polyneuropathy (CIDP) is the most common acquired chronic

Figure 1. Guillain-Barre syndrome disability score changes in time in patients with prednisone therapy.

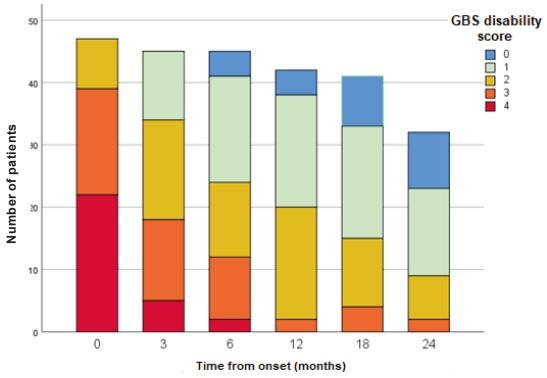
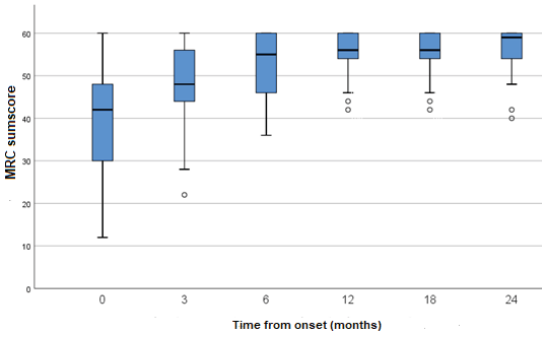


Figure 2. Medical Research Council score changes in time in patients with prednisone therapy.



inflammatory neuropathy [9]. Some studies report a male predominance and age varies between 30 to 60 years [2,10].

The most common comorbidity in our population was diabetes. At the present time there is controversy about CIDP association with diabetes. In the past, epidemiological studies reported that CIDP was nine times more frequent in patients with previous diagnosis of diabetes. In world reports, 9% of CIDP patients have diabetes. In our population, this frequency was higher (19.1%), which may be related to a higher rates of obesity, metabolic syndrome and diabetes in our country [11,12].

Cranial nerve involvement was uncommon, being the facial nerve the most frequent affected in our series, similar to other series [2]. Acute-onset CDIP presented in 18%, similar to previous reports [13,14]. Absence of cranial nerves involvement, and no need for mechanical ventilation should raise suspicion for acute-onset CIDP. In our study, 20% had cranial involvement and none required mechanical ventilation [15].

Atypical variants were found in 49% in other series. Nonetheless, in our study the frequency found was 21.2% [3]. Atypical variants included tremor-ataxia related to neurofascin-155 (NF155) antibodies, contactin 1 (CNTN1) and contactin-associated protein 1 (Caspr1), unfortunately in our population we did not perform antiganglioside testing [16]. The pure-motor variant has been classically linked to poor steroid therapy response and with excellent response to IVIg. None of our patients had a pure-motor variant. Time from symptom onset to diagnosis in patients with atypical variants was similar in our patients compared with other reports [17]. Additionally, other studies report infrequent CSF protein-dissociation in atypical CIDP. However, our results differ from those reported in other series [2].

Treatment for CIDP is well standardized, including steroid therapy and IVIg as maintenance [6]. Factors associated to good response have not been described for any particular therapy [1]. Decision to start and continue treatment is at discretion of the physician, considering many variables, including costs. In first world countries there is a greater access to IVIg and SCIG, while in Mexico this therapy is not easily accessible for patients. Therefore, steroids remain the cornerstone for CIDP treatment in such countries.

CIDP is classically a chronic inflammatory steroid-responsive polyneuropathy. Different steroid regimens have been used, such as intravenous methylprednisolone pulses of 500-1,000 mg for five days monthly, dexamethasone 40 mg orally each day for four days every month and prednisone 1-2 mg/kg daily with gradual dose decrease, without differences in the clinical response with those different schemes [7]. In our experience, we started treatment with prednisone at 50 mg as a standard dose in all patients, gradually decreasing each month prior to clinical evaluation, to avoid side effects at higher doses. Adverse effects to prednisone treatment were reported in 40.4%, being with weight gain /obesity being the most frequent.

Several scales have been used to assess the clinical response to CIDP therapy. A retrospective study used the Rankin scale, reporting that 60% of the patients are steroid responders [18]. In other clinical trial or retrospective studies the INCAT scale is

Table III. Clinical response at follow-up.

	Initial *n* = 47	3 months *n* = 46	6 months *n* = 44	12 months *n* = 42	18 months *n* = 41	24 months *n* = 32	*p* value
GBS disability score, median (IQR)	3 (2.25-4)	2 (1.25-3)	2 (1-2)	1.5 (1-2)	1 (1-2)	1 (0-2)	< 0.001
MRC score, mean, SD	39.5 ± 12	47.7 ± 9.5	50.5 ± 11.5	54.9 ± 5.1	55.1 ± 5.7	56.3 ± 5.1	< 0.001
Independent gait (%)	8 (17%)	27 (60%)	34 (75.6%)	39 (92.9%)	37 (90.2%)	30 (93.8%)	< 0.001
Prednisone dose (mg), median (IQR)	50 (32.5-50)	40 (20-40)	30 (20-40)	10 (5-20)	5 (0-10)	1 (0-5)	< 0.001

GBS: Guillain-Barre Disability Score; IQR: interquartile range; MRC: Medical Research Council; SD: standard deviation.

used, observing response to treatment from weeks 6 to 9 from the start of therapy. We decided to use GBS disability score as reported in other clinical trials [19,20].

In our population, 63% presented clinical improvement (decrease of at least one point on Guillain-Barré Disability Score) at three months since therapy initiation. Interestingly, we did not observe a significant difference between patients who received steroid + immunotherapy (IVIg or PE) at diagnosis. A delay in CIDP diagnosis implies delaying the initiation of immunotherapy to decrease peripheral nerve damage, therefore poor clinical response. Previous studies concluded that patients with a delay > 5 months have greater weakness in muscle strength and greater disability at diagnosis, in addition to less short and long-term disease control [21]. In our patients we observed similar results. Patients with time since symptom onset to diagnosis a ≥ 6 month, did not had a favorable clinical response at 3 months. Steroid therapy has shown efficacy in the treatment of CIDP in the short and long term. Our study shows significant results in improvement and disease control at two years of follow-up. Eighty-percent of patients at diagnosis had lost independent gait, and after steroid therapy we observed an increase in patients who regained independent gait. Only 6.3% continued without independent gait at one year follow-up [20]. Previous clinical trials reported that 50% of patients treated with prednisone presented disease relapses at 11-month follow-up (median 5-63). Our results show similar conclusions [22]. The use of steroid-sparing therapy is still controversial. The recommended agent nowadays is azathioprine. We prefer to start treatment with prednisone only and in cases that a more rapid decrease steroid is needed, as in patients with diabetes or those with early side effects, we use steroid-sparing therapy. Forty-five percent of our patients received azathioprine. Patients with atypical variants were initially treated with IVIg and prednisone, as well as cyclophosphamide completing 9 pulse of 1 g. This therapy demonstrated also a good outcome, as mentioned previously [23].

One of the limitations of the study is its retrospective nature. However, our findings support prednisone treatment in CIDP patients who do not have access to IVIg. Another limitation was that the INCAT score was not calculated due to the retrospective nature of our investigation.

Conclusion

Steroid therapy is an appropriate option in patients with CIDP in low-income countries. Delay in diagnosis and atypical variants are characteristics related to poor clinical response to prednisone therapy. This study provide background for further prospective studies in Latin America.

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Eficacia del uso de prednisona como terapia a largo plazo en pacientes con polineuropatía desmielinizante inflamatoria crónica (PDIC): una cohorte retrospectiva

Introducción. Los pacientes con polineuropatía desmielinizante inflamatoria crónica (PDIC) responden adecuadamente a la terapia con esteroides y a la inmunoglobulina intravenosa (IgIV). Sin embargo, pocos pacientes tienen acceso a la IgIV en los países en desarrollo. Existe poca información sobre la respuesta clínica a la terapia con esteroides en los países de Latinoamérica.

Objetivo. Describir la respuesta clínica funcional a largo plazo (24 meses) a la terapia con prednisona en pacientes con PDIC.

Material y métodos. Se realizó una cohorte retrospectiva. La selección incluyó a pacientes con diagnóstico definitivo de PDIC según los criterios europeos de la Clínica de Enfermedades Neuromusculares del Instituto Nacional de Neurología y Neurocirugía entre enero de 2016 y diciembre de 2020. La buena respuesta a la terapia con esteroides se definió como una mejoría al menos en un punto de la *Guillain-Barre Disability Score* (GBS). La mala respuesta a la terapia con esteroides se definió como pacientes que no mostraron mejoría al menos en un punto en la GBS. Los pacientes fueron evaluados a los 3, 6, 12, 18 y 24 meses.

Resultados. Se incluyó a 47 pacientes con PDIC. La mitad de ellos eran varones y la edad media fue de 46 ± 15 años. El tiempo medio desde el inicio de los síntomas hasta el diagnóstico fue de 6 (rango intercuartílico: 2-12) meses. La variante clínica más común fue la sensomotora (57,4%), seguida de la PDIC de inicio agudo (21,3%) y de variantes atípicas (21,2%). En el momento del diagnóstico, nuestros pacientes presentaban: GBS media de 3 (2,25-4) puntos, puntuación de la escala del Medical Research Council (MRC) de 39,5 ± 12 puntos, marcha independiente en el 17% y dosis media de prednisona

de 50 mg (32,5-50). Veinticuatro meses después de la terapia con prednisona, la GBS media era menor –1 (0-2) puntos–, la puntuación media del MRC era de 56,3 ± 5,1 puntos, había marcha independiente en el 93% y la dosis de prednisona era de 1 mg (0-5). Los pacientes con mala respuesta clínica funcional a los tres meses tuvieron un retraso en el diagnóstico > 6 meses (64,7% frente a 27,5%) y variantes clínicas atípicas (47% frente a 6,8%).

Conclusión. Los pacientes con PDIC tratados con prednisona tienen una buena respuesta clínica funcional a largo plazo. El retraso en el diagnóstico y la variante atípica son características clínicas frecuentes de la respuesta clínica funcional deficiente en el tratamiento con prednisona.

Palabras clave. Crónica. Neuropatía. PDIC. Prednisona. Pronóstico. Tratamiento.