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Short communication

# Epidemiology of chronic inflammatory demyelinating polyneuropathy in the South-Eastern area of Santiago, Chile

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ABSTRACT

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated polyneuropathy. It usually has an insidious onset, progressive course and heterogeneous clinical features. As far as we know, there is no epidemiological information on CIDP in South America and the Caribbean. Our aim was to estimate the frequency of CIDP in the South-Eastern region of Santiago, where our hospital is based and the population number assigned is officially reported every year by the health authorities.

Records of 581 patients registered with the diagnosis of neuropathy were found and all patients meeting the diagnostic criteria of the EFNS/PNS for definitive and possible CIDP were included. Data were collected using a data extraction protocol designed by the authors and which included demographic, clinical, laboratory and electrophysiological information.

The estimated prevalence and incidence of CIDP were 2.95/100,000 and 0.46/100,000 respectively. Fifteen patients (8 men, 7 women) were classified as definitive or possible CIDP. Nine patients had typical CIDP and three also had diabetes mellitus. The prevalence and incidence rates were similar to those reported in other regions of the world.



## 1. Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune-mediated disorder of the peripheral nerves and nerve roots causing limb weakness and sensory deficits [1]. In a recent *meta*-analysis that includes patients from Europe, USA, Japan and Australia, the pooled crude incidence rate of CIDP was estimated in 0.33 per 100,000 person-years and the prevalence rate in 2.81 per 100,000 [2]. In Chile there is epidemiological information on the incidence of acute inflammatory demyelinating polyradiculoneuropathy [3]. However, as far as we are aware, there is no epidemiological information on CIDP in South America and the Caribbean.

In Chile, 78% of the population receives health care through a publicly funded health care system (FONASA) [4]. Santiago has a population of 6.179.183 inhabitants which is 35.2% of the total population of Chile. Our hospital, Hospital del Salvador, is located in the South-Eastern region of Santiago and has an assigned population of 507,745 people ≥15 years of age, with FONASA insurance. In this area, we conducted a population based retrospective study to estimate the incidence and prevalence of CIDP. We hope that this work contributes to close the information gap on CIPD in our region.

## 2. Methods

We used the hospital database that contains information on all admissions and outpatient visits. Patients included in our study had to be alive and reside in the catchment area on the prevalence day. Patients living outside the catchment area were excluded.

We searched for all medical records with the following diagnostic codes according to the International Classification of Disease, 9th Revision, Clinical Modification (ICD9-CM): acute polyneuritis (357.0), polyneuropathy in other diseases (357.4), other neuropathy (357.8), unspecified neuropathy (357.9) [5]. Additionally, in order to have an independent source of information, we

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interviewed all 24 neurologists working in our hospital, and requested information on any patients with CIDP or acquired demyelinating neuropathy.

The diagnostic criteria of the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) for definite and possible CIDP were used [6]. Patients having concomitant diabetes were included. We reviewed the national identification number of all patients in the Chilean Identification Service to check if they were alive on the first of April 2019.

We reviewed the records of all included patients using a standardized format to collect information in an anonymized database that included demographic, clinical, laboratory and electrophysiological data. Additionally all patients were evaluated by the authors to confirm findings.

### 2.1. Statistical analysis

Demographic data were obtained on the population $\geq$ 15 years of age assigned to the hospital. Estimated mean incidence was calculated, with the 3 years prior to the prevalence date (1 May 2018). General crude prevalence and incidence rates, as well as the breakdown per gender and age were determined. We calculated 95% confidence intervals (CI) assuming a Poisson distribution and the Student's *t*-test was used to compare means.

The study protocol received ethics approval from our local Research Ethics Committee.

## 3. Results

We reviewed a total of 581 charts during the 2-year study period. Fig. 1 shows the selection process. Fifteen cases (8 males, female 7; 1.1 ratio) fulfilled the EFNS/PNS criteria for definitive or possible CIDP and all were alive on the prevalence day. The mean incidence rate was 0.46 (95% CI 0.00–1.05) and prevalence was 2.95 (95% CI 1.46–4.45) per 100,000. Mean age at onset was 49.8 (SD 13.1) years (range 17–73), men 55.25 years (SD 7.19), women 43.57 years (SD 15). See Table 1 for details.

## 4. Discussion and conclusion

Our estimated incidence rate was 0.46 (95% CI 0.00–1.05) and prevalence of CIDP was 2.95 per 100,000 (95% 1.45–4.45), figures that are within the range of those reported in the European, Australian and Japanese populations [7–11]. Our figures were obtained from a well defined, complete storage system of patient records and we also surveyed all the neurologists working in the hospital in order to detect all cases of definitive or possible CIDP. Clinical data were extracted and classified according to a protocol that was defined prior to the data collection and patients were evaluated by the authors. Chile has a reliable national identity number database, so we were able to reduce survival bias by checking the national database of births and deaths. We did not include patients under 15 years old. However, CIDP is extremely rare in people under 15 years of age [12], so this probably does not affect our data.

This study may have selection bias, because we used retrospective collect data. We tried to minimize this by reviewing the records of all patients recorded as having a neuropathy. CIDP does not have a diagnostic gold standard but this is a problem with all CIDP studies. We decided to use EFNS/PNS criteria as these are more sensitive than other clinical criteria.

We found in our group a male to female ratio of 1.1:1, but most series showed a slightly greater difference with a male predominance. Our data might have sample bias or represent our particularly population tendency.

Chile has the highest prevalence of diabetes in South America with 9,4% of the adult population affected. [13]. We found 3 diabetics in 15 CIDP patients and a crude prevalence of 0.59 per 100.000 opposed to 8 $\times$ 100.000 in the work of Dyck [14]. Therefore we can not establish an association between them.

Finally, as far as we know this is the first attempt to rigorously produce an epidemiological estimation of CIDP in South America and the Caribbean. We are aware that CIDP has low incidence and that it is necessary in the future to evaluate larger populations in multicentric studies.

**Table 1**
Mean Incidence rate, and crude prevalence rates (per 100,000 population) by age group and associated with diabetes.

| Incidence rate (/100,000) | | Rates | (95% CI) |
|---|---|---|---|
| Mean (3 yrs) | 4 | 0.46 per 100,000 | (0.00–1.05) |
| Prevalence rate (/100,000) | n | Rates | (95% CI) |
| Young adult 15–55 yrs | 10 | 1.97 per 100,000 | (0.75–3–19) |
| Elderly adult 55+ yrs | 5 | 0.98 per 100,000 | (0.12–1.85) |
| CIDP associated with diabetes | 3 | 0.59 per 100,000 | (0.00–1.26) |
| Crude global prevalence | 15 | 2.95 per 100,000 | (1.46–4.45) |

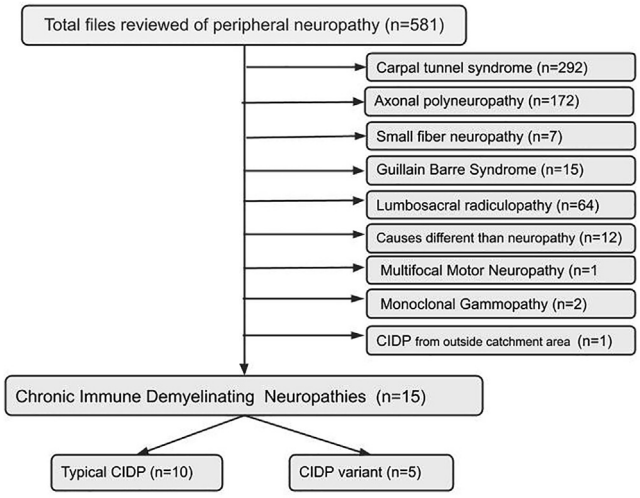


**Fig. 1.** Flowchart of inclusion cases.

## Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

## Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jocn.2020.02.009.

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