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Research Article

# Clinical and Electrophysiological Profile of Patients with Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) in a Tertiary Care Centre

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## Abstract

**Introduction:** Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is an acquired immune-mediated peripheral neuropathy. This study was undertaken to describe the clinical and electrophysiological profile of patients with CIDP from a single centre.

**Aims and Objectives:** 1. To study the clinical and electrophysiological profile of patients with CIDP. 2. To study the effects of treatment and their outcomes in patients with CIDP through the inflammatory neuropathy cause and treatment (INCAT) scoring system and INCAT overall disability sum score (ODSS).

**Materials and Methods:** It is an ambispective study conducted between July 2017 and December 2019 in the Department of Neurology, Nizam's Institute of Medical Sciences. A structured proforma and nerve conduction studies were done.

**Results:** The mean age was 41.32 years. Male: Female = 38:12. Twenty-four patients had a relapsing remitting course, 24 patients had a progressive course, and 2 had an acute onset of Guillain-Barré syndrome presentation. Typical CIDP was seen in 42 (84%) patients, 5(10%) patients had Multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) variant, 2 patients had pure motor variant, 1 patient had pure sensory variant, and none of them showed distal acquired demyelinating symmetric (DADS) phenotype. Slow conduction velocity was the most common observation seen in 94% patients.

**Conclusions:** Typical CIDP was the most common phenotype, and none of the subjects had the DADS variant; thus, we can hypothesise that the prevalence of DADS in South India is less compared to the Western studies.

**Key message:** CIDP is an immune-mediated disorder with various phenotypes. Treatment should be tailored made depending upon the phenotype. This is one of the largest cohorts from a single centre in South India.

***Keywords:*** *Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP), European Federation of Neurological Societies/ Peripheral Nerve Society (EFNS/PNS) criteria, Multifocal Acquired Demyelinating Sensory and motor neuropathy (MADSAM)*

## Introduction

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is an acquired peripheral neuropathy resulting from an autoimmune attack on peripheral nerve myelin [1]. Dyck et al introduced the name chronic inflammatory polyradiculoneuropathy [2]. The reported prevalence of CIDP ranges from 0.7 to 10.3 cases per 100,000 people [3]. The male-to-female ratio of 1.5 – 4:1 [3]. We intend to study the clinical and electrophysiological profile and outcome measures of a South Indian cohort from a single centre.

**Pathogenesis:**

It has been proposed that both cell-mediated and humoral mechanisms act synergistically in the pathogenesis of CIDP [4].

During active disease CD4+ T cells in the periphery upregulate activation markers such as t-bet and pstat1 [5] and secrete proinflammatory cytokines including interleukin (IL)-2, interferon γ (IFNγ) and IL-17 as well as the chemokines [6], interferon gamma-induced protein (IP)-10 and macrophage inflammatory protein 3 β (MIP3β) which induces up regulation of vascular cell adhesion molecule (VCAM)-1[7], endothelial leukocyte adhesion molecule (ELAM)-1[8]and intercellular adhesion molecule (ICAM)-1[9] on endothelial cells lining the blood vessels of the nerve. In CIDP, sural nerve biopsies show the infiltrating inflammatory cells [10]. Immunoglobulin and complement can be seen deposited on the outer surface of Schwann cells of the compact myelin in the sural nerve biopsies [11].

Axoglial proteins are important for the formation and maintenance of the nodes of Ranvier and the paranodal regions of myelinated axons [12]. The nodal cell adhesion molecules (CAMs), gliomedin, neuron glia-related CAM (NrCAM), and neurofascin 186 (NF186) are vital for the initial clustering of Na+ channels during development [12]. The adjacent paranode consists of axoglial junctions between paranodal loops and axonal membrane composed of contactin-1/ caspr-1 complexes, which bind to Schwann cell neurofascin 155 (NF155) [13]. NF155 is essential for ion channel segregation, paranodal structure and efficient nerve conduction [14]. Antibodies against the CAM neurofascin have been identified in 4% of patients with CIDP, mainly the glial neurofascin isoform NF155 [15].

**Aims and Objectives**

1. The primary objective is to study the clinical and electrophysiological profile of patients with CIDP.

2. To stratify the patients with CIDP into various clinical phenotypes and to group the patients into definite, probable and possible categories according to the European Federation of Neurological Societies/ Peripheral Nerve Society (EFNS/ PNS) criteria.

3. To study the effects of treatment options and their outcomes in patients with CIDP through the inflammatory neuropathy cause and treatment (INCAT) scoring system and INCAT overall disability sum score (ODSS).

## Materials and Methods

It is an ambispective observational study conducted between July 2017 and December 2019 in the Department of Neurology, Nizam's Institute of Medical Sciences, Hyderabad. Approval from the ethics committee was obtained.

**Inclusion criteria:**

1) All the patients diagnosed to have CIDP by clinical, electrophysiological EFNS/PNS criteria were included in the study.

2) All the patients who have given consent to participate in the study.

**Exclusion criteria:**

1) Patients with confirmed mimics.

All the consecutive patients with clinical characteristics of CIDP were taken from the registry. They have undergone a direct structured interview using a pre-formatted standard proforma. All the patients were evaluated and stratified by the treating Neurophysician. The diagnosis of CIDP was based on EFNS/PNS CIDP criteria [16]. Nerve conduction studies were performed using a Natus Synergy electromyography machine. Motor nerve studies were made of the median, ulnar, peroneal and tibial nerves, including F wave analyses.

Demyelinating features like prolonged latencies (≥50% of normal), conduction velocity slowing (≤70% of normal), abnormally prolonged F waves (≥130% of normal), and temporal dispersion were studied for all the motor nerves. Partial motor conduction block was defined as a definite >50% and probable >30% reduction of compound muscle action potentials between the stimulus sites. Antidromic sensory nerve conduction studies were performed in the median and the sural nerves. Supportive criteria included analysis of CSF for albumino-cytological dissociation, MR neurography and nerve biopsy. INCAT ODSS [18] and MRC sum scoring were used to assess the functional disability.

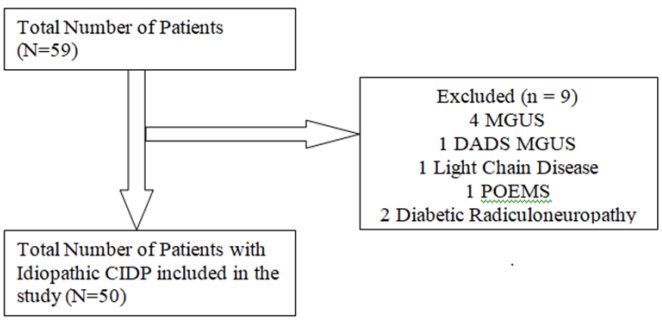
A total of fifty-nine patients were assessed (Figure 1). Among them, nine patients were excluded due to secondary diagnosis of either paraproteinemic neuropathy or diabetic radiculoneuropathy. The remaining fifty patients fulfilling EFNS/PNS criteria were included in the study and classified into the relapsing remitting (RR) and non-relapsing remitting group (Non-RR). Patients who had definite relapses without complete recovery were in the relapsing group. Cases with a steadily progressive course, and those with a sub-acute onset and monophasic course have been included in the non-relapsing group.

**Statistical Analysis**

Descriptive analysis was carried out by mean and standard deviation for quantitative variables, frequency and proportion for categorical variables. Data was also represented using appropriate diagrams like a bar diagram and, pie diagram. Data were entered into an electronic database for statistical analysis (SPSS, version 20.0). Data were presented as numbers (percentages) or means (standard deviation [SD]) as appropriate. Categorical variables were compared using the chi-square test or Fisher's exact test, and continuous variables using the Student t test for independent samples. Kaplan-Meier analysis was used to illustrate the time to diagnosis, and the Log-Rank test was used to compare the means of time to diagnosis. The p-value of less than 0.05 was considered statistically significant.

## Results

The mean age of presentation was 41.32±3.4 years. Among 50 patients, 38 (76%) were males and 12 (24%) were females. In the RR group (52%) mean age was 42.38 years, and the mean age in the non-RR group was 40.17 years. Out of the 50 patients, 2 patients (4%) had Guillian-Barré Syndrome (GBS) to CIDP progression (qualifying to A-CIDP with respect to duration of progression >8 weeks), 24 patients (48%) had a relapsing remitting course, and 24 (48%) patients had a progressive course. A maximum of ten relapses were observed in one female patient over a span of 2 years. She had relapses on tapering immunomodulation during pregnancy and had relapses even in the postpartum period. Her illness was responsive to IVIG. The mean number of relapses was 1.48 ±2.013(SD). Typical CIDP was seen in 42 (84%) patients, and 8 (16%) patients had CIDP variants (Figure 2). This was strictly based on the EFNS/PNS 2010 criteria. It was observed that the mean time to diagnose typical CIDP was 4.2 months compared to 18.188 months in CIDP variants. (significant p value 0.000). (Figure 3)




***Figures 1.*** *Sample Size and subjects recruited in the study.*

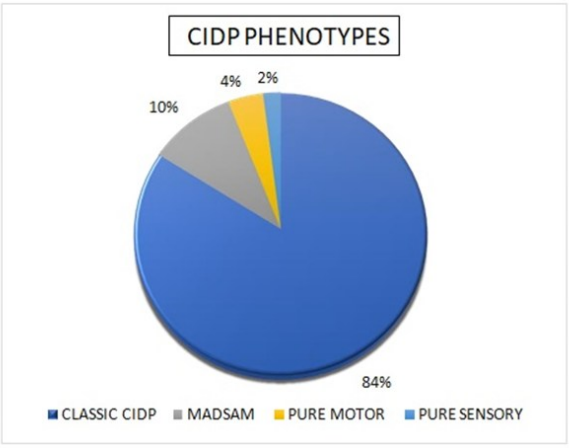



***Figure 2.*** *Pie chart distribution of clinical phenotypes*

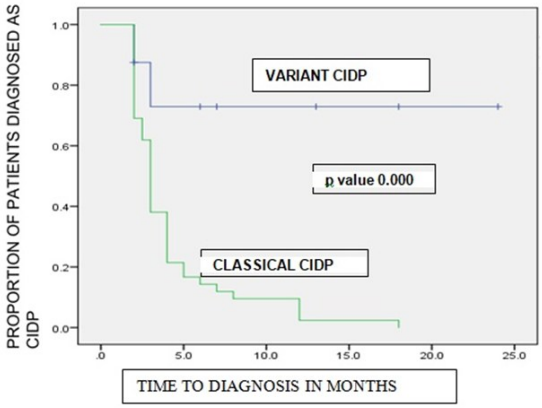



***Figure 3.*** *Kaplan Meier curve to compare time to diagnosis in classical CIDP and variant CIDP patients*

Clinical features and lab features in the study population have been depicted in Table 1.

**Table 1.** Distribution of clinical manifestations and lab features.

| | Clinical and lab feature | Number (n) | Percentage |
|---|---|---|---|
| Motor symptoms and signs | Hypo or areflexia | 50 (50) | 100% |
| | Symmetrical | 46 (50) | 92% |
| | Proximal ≥ Distal | 34 (50) | 68% |
| | Distal ≥ Proximal | 15 (50) | 30% |
| | Muscle atrophy | 21 (50) | 42% |
| Sensory symptoms | Numbness | 44 (50) | 88% |
| | Sensory ataxia | 23 (50) | 46% |
| | Paraesthesias | 41(50) | 82% |
| | Burning pains | 12 (50) | 24% |
| | Tremor | 8 (50) | 16% |
| | Pure sensory symptoms withoutweakness | 1 (50) | 2% |
| Cranial nerve symptoms | Facial | 15 (50) | 30% |
| | Ophthalmoparesis without ptosis | 1 (50) | 2% |
| | Ptosis | 1 (50) | 2% |
| | Bulbar | 0 (50) | 0 |
| | Autonomic manifestations | 3 (50) | 6% |
| | Respiratory failure | 3 (50) | 6% |
| Lab features | Nerve Biopsy Total | 14 | |
| | Nerve Biopsy Abnormal | 12 (14) | 86% |
| | Nerve Biopsy Normal | 2 (14) | 14% |
| | HRUS Total | 9 | |
| | HRUS Abnormal | 8 (9) | 89% |
| | HRUS Normal | 1 (9) | 11% |
| | MRN Total | 28 | |
| | MRN Abnormal | 23 (28) | 82% |
| | MRN Normal | 5 (28) | 16% |

*HRUS: High resolution Ultra Sound, MRN: Magnetic Resonance Neurography*

As per the hospital protocol, nerve biopsy was performed for all the patients with a diagnosis of probable or possible CIDP. Fourteen patients underwent nerve biopsy.

"Definite criteria" were met by 88% (44/50) of patients, and 12% (6/50) met the "Probable criteria". The electrophysiological parameters have been elicited in Figure 4. Slow conduction velocity was the most common observation in our study.

Lumbar puncture was performed in 33 patients. Out of which 24 (73%) patients showed albumino cytological dissociation (ACD). A cell count of less than 10 cells was seen in 32 patients. The mean CSF glucose levels were 88.67±6.5 mg/dl. The mean CSF protein was 133.42±7.4 mg/dl. None of the patients had any evidence of human immunodeficiency virus (HIV), Lyme's disease, root infiltration or other autoimmune disorders.

Treatment modalities used in the study cohort are shown in Figure 5. The choice of therapy was based on the clinician's discretion and feasibility.

INCAT ODSS and MRC grading were used for disability assessment in CIDP patients. (Table 2) The mean time for follow-up of CIDP patients in this study was 1.5 years (1 to 3.5 years). Patient outcome was categorised as worsened, stable or improved and was assessed at 6 months and one year based on the CIDP disease activity scale (CDAS). At 6 months of follow-up 46% had worsened, 16% had improved, 38% remained stable, and at one year of follow up none of them had worsened, 20% had improved completely, 80% remained stable. Mean INCAT ODSS score at onset, 6 months follow-up up and 1 year follow-up was 6.46, 5.40 and 3.30, respectively (p value = 0.000). (Table 3)

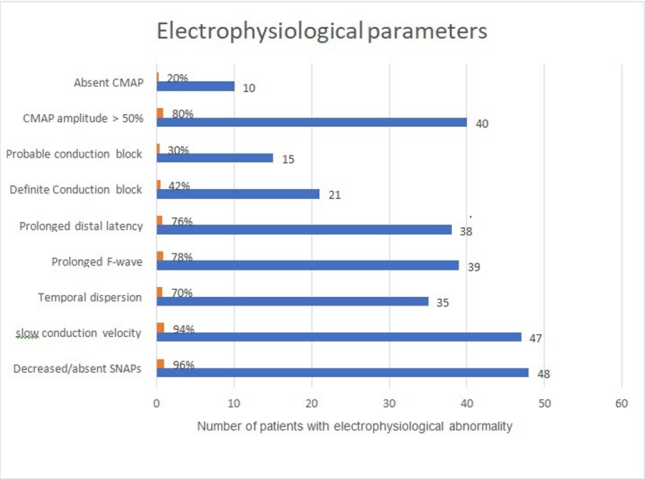



***Figure 4.*** *Distribution of nerve conduction parameters in the cohort – electrophysiology data.*

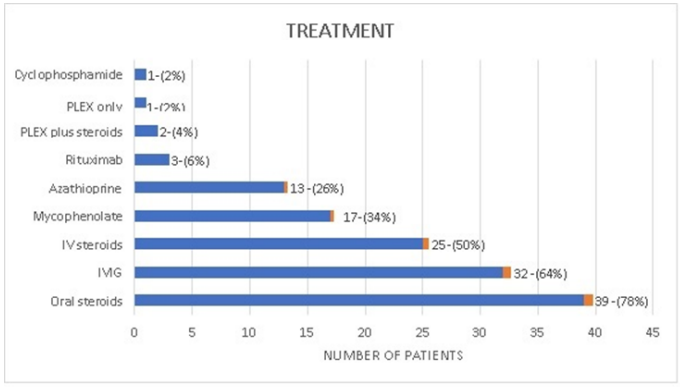



***Figure 5.*** *Treatment details in the cohort with percentages.*

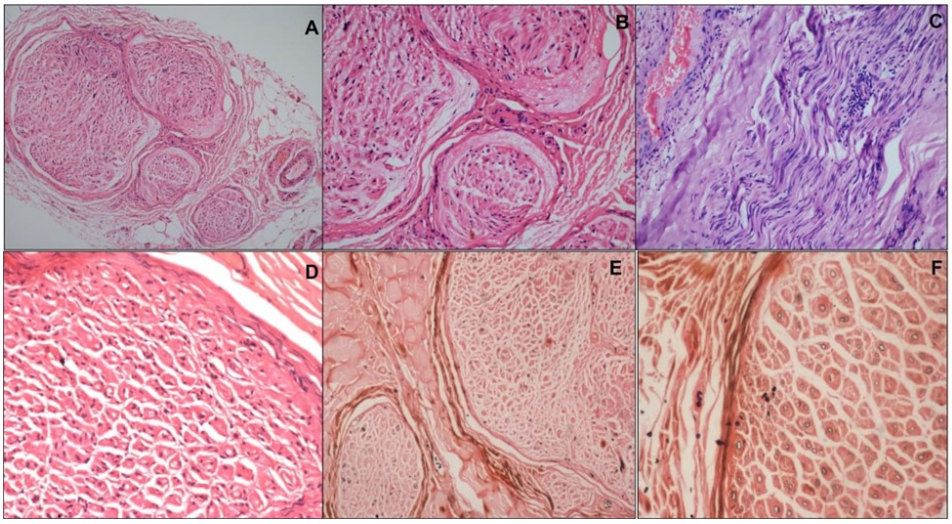



***Figure 6.*** *Nerve biopsy in CIDP shows onion bulb patterns, segmental demyelination and inflammatory infiltrates.*

**Table 2:** INCAT ODSS and MRC GRADING Pre and Post treatment.

| | Minimum Score | Maximum score | Mean | SD | N | p value |
|---|---|---|---|---|---|---|
| Pre treatment INCAT | 2 | 12 | 6.46 | 2.409 | 50 | 0.000 |
| Post treatmentINCAT | 0 | 8 | 3.30 | 1.568 | 50 | |
| Pre treatment MRC Score | 0 | 60 | 44.12 | 10.898 | 50 | 0.000 |
| Post treatment MRC Score | 36 | 60 | 52.00 | 6.178 | 50 | |

***INCAT:*** *Inflammatory neuropathy cause and treatment, MRC: Medical Research Council, ODSS: Overall Disability Sum Score, SD: Standard Deviation*

**Table 3.** Comparison of means of INCAT scores at onset and 6 months and one year of follow-up.

| CIDP criteria | Sensitivity (%) |
|---|---|
| AAN | 11% to 63% |
| Saperstein et al. | 47% to 70% |
| Hughes/INCAT | 43% to 80% |
| Nicholas/modified INCAT | 50% to 95% |
| EFNS/PNS | 81% |
| Koski et al. | 63% |

*AAN: American Academy of Neurology; CIDP: Chronic Inflammatory Demyelinating Polyradiculoneuropathy; EFNS: European Federation of Neurological Societies; PNS: Peripheral Nerve Society; INCAT: Inflammatory neuropathy cause and treatment*

## Discussion

CIDP is one of the commonly encountered immune-mediated neuropathies in clinical practice with a wide variety of clinical presentations as cited below:

Typical CIDP exhibit a slowly progressive course, but a relapsing-remitting course is noted in at least one-third and may be more common in younger patients [16]. Typical CIDP is predominantly symmetric sensorimotor polyneuropathy with motor involvement exceeding sensory involvement [19].

Joint Task Force-Second revision report was released in 2021 by the European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) to revise the 2010 consensus guideline on CIDP [20]. According to this revision, "the previous term 'atypical CIDP' was replaced by 'CIDP variants' because these are well-characterised entities (multifocal, focal, distal, motor, or sensory CIDP). The levels of diagnostic certainty were reduced from three (definite, probable, and possible CIDP) to only two (CIDP and possible CIDP), because the diagnostic accuracy of the criteria for probable and definite CIDP did not significantly differ. Good Practice Points were formulated for supportive criteria and investigations to be considered to diagnose CIDP. The principal treatment recommendations were: (a) intravenous immunoglobulin (IVIg) or corticosteroids are strongly recommended as initial treatment in typical CIDP and CIDP variants; (b) plasma exchange (PLEX) is strongly recommended if IVIg and corticosteroids are ineffective; (c) IVIg should be considered as first-line treatment in motor CIDP (Good Practice Point); (d) for maintenance treatment, IVIg, subcutaneous immunoglobulin or corticosteroids are recommended; (e) if the maintenance dose of any of these is high, consider either combination treatments or adding an immunosuppressant or immunomodulatory drug (Good Practice Point); and (f) if pain is present, consider drugs against neuropathic pain and multidisciplinary management (Good Practice Point) [20].

An extended protocol for nerve conduction studies has been proposed by the joint task force, which includes four motor and four sensory nerves [20]. Since we conducted this study before the release of the second revision report of EAN/PNS, the results from our study are as per the 2010 EFNS/PNS guidelines.

**Atypical CIDP variants:**

Sensory predominant CIDP: It occurs in 5–35% of patients, often starting with lower limb numbness [21]. Despite purely sensory symptoms, patients often demonstrate prominent motor nerve conduction abnormalities consistent with demyelination [21]. A subset of patients with CIDP (~5%) present with progressive sensory ataxia and sensory symptoms, termed chronic immune sensory polyradiculopathy (CISP) [22]. However, according to the EAN/PNS guidelines, CISP is not considered a variant of CIDP [20]. Our study showed a small percentage (2%) of subjects with the pure sensory phenotype.

Distal acquired demyelinating symmetric neuropathy (DADS): DADS refers to a distal and sensory-predominant variant of CIDP [23]. In 50–70% of patients, an IgM paraprotein having anti-myelin-associated glycoprotein (anti-MAG) antibody activity is responsible for the pathogenesis [23]. It is a surprising observation that our centre has only one reported case of DADS in the last 4 years, and none in the current study had DADS.

Motor dominant CIDP (Pure motor variant): It has been reported in 7–10% of patients with CIDP, especially in those younger than 20 years [24,25,26]. Our study had 4% of patients with this variant of CIDP.

Lewis-Sumner syndrome (LSS) or multifocal acquired demyelinating sensory and motor neuropathy (MADSAM): It is characterised by asymmetry, presenting as a multifocal multiple mononeuropathy most commonly in the upper limbs. It accounts for 6–15% of CIDP patients [27]. Aligning with the existing literature, we had 10% of our study subjects with the MADSAM phenotype. This was the most common phenotype following the typical CIDP in our study.

Contactin 1 antibody-mediated: The clinical phenotype of contactin 1 antibody-mediated CIDP is not fully established but is often severe and predominantly motor with early axonal involvement [13]. Patients with antibodies against Neurofascin and Contactin 1 do not typically respond to intravenous immunoglobulin (IVIG) but can be responsive to B-cell depletion therapy.

These antibodies have not been tested in our cohort, as they are commercially unavailable at our centre.

Multiple sets of diagnostic criteria have been published for the diagnosis of CIDP (Table 4) [16,17,28]. Corresponding to the other studies, our study had slowing of the conduction velocity as the most common electrophysiological observation, followed by prolongation of F wave latency, followed by prolonged distal motor latency.

Another interesting observation was decreased SNAPs, which could probably indicate severe secondary axonal damage. An interesting observation in our study was the presence of cranial neuropathy (ptosis and ophthalmoparesis) in patients with typical CIDP.

The diagnostic utility of nerve biopsy (typically of the sural nerve) for suspected CIDP is controversial [29]. Nerve biopsy is used mainly when other studies fail to clearly establish the diagnosis of CIDP. As per the institute protocol, nerve biopsy was performed in the patients with probable or possible CIDP after informed consent. We performed a nerve biopsy in 14 subjects. Nerve biopsy in CIDP shows onion bulb patterns, segmental demyelination and inflammatory infiltrates (Figure 6). The presence of macrophage clusters (three or more macrophages around endoneurial vessels) in sural nerve biopsies may serve as a useful additional marker for establishing the pathologic diagnosis of CIDP [29].

The primary goals of the treatment for CIDP are to reduce symptoms, improve functional status and maintain long-term remission. The widely accepted first-line therapy in CIDP consists of corticosteroids, IVIg, and PLEX [30]. Improvement can be expected in 50–80% of the patients [30]. There were some interesting observations with respect to the treatment in our case series. One patient with the maximum number of relapses (10 in 2.5 years) showed a dramatic response to IVIg and had a relapse each time IVIg was stopped. Currently, this patient is being maintained on 5 grams of IVIg every 75 days and is in complete remission. Three pediatric patients (<16 years) were given IVIg, and all of them showed near complete improvement in disability. The youngest patient in our cohort (a 3-year-old child) is in complete remission after stopping treatment at 2 years of disease onset.

When diabetic and non-diabetic groups were compared, there was no difference between the groups, similar to a study by Dunnigan SK et al [31].

We found a non-RR disease course (52%) to be more common than RR, which is similar to or higher than the other studies (Iceland (76%), [32] Italy (74%), [33] Olmsted County (56%) [34] and England (48%) [35].

This is one of the large cohort studies of CIDP subjects from a single tertiary care centre in South India reflecting the clinical and electrophysiological profile.

## Limitations

Enrolment from a single-center, as NIMS is a tertiary care center, most patients enrolled are of high disability thus with unintentional selection bias.

Antibodies implicated in autoimmune nodopathies not tested.

## Conclusion

Currently the concept of CIDP includes several clinical subtypes, typical CIDP and other variants. Typical CIDP was the most common phenotype and none of the subjects had DADS variant, thus we can hypothesize that the prevalence of DADS in South India is less compared to the Western studies. The conduction velocity slowing was the most common electrophysiological abnormality in our study. Larger studies are warranted with special attention to diagnosis, the expanding CIDP spectrum, recognition of recently discovered antibodies and newer treatment advances.

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