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Clinical and Electrophysiological Profiles of Chronic Inflammatory Demyelinating Polyradiculoneuropathy: Experience at Referral Neuroscience Hospital in Bangladesh

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Abstract

Background: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare and heterogeneous immune-mediated neuropathy. **Objectives:** The aim of this study was to evaluate the clinical and electrophysiological aspects of CIDP in adult. **Methodology:** This cross-sectional study was carried out in the department of Neurophysiology of National Institute of Neurosciences and Hospital, Bangladesh from July 2020 to July 2022. We included 50 consecutive patients aged 18 to 70 years fulfilling criteria for CIDP proposed by the European Federation of Neurological Societies and the peripheral nerve society, 2010. These patients were evaluated by detailed history, physical examination and electrophysiological findings. **Results:** The mean age was 43.82 ± 13.43years. Males were more affected than females. Weakness and paresthesia were the most common symptoms. Forty-six patients (92%) had a progressive and four patients (8%) had a relapsing course. Most patients were classified as typical CIDP (90.0%) and forty-six patients (92.0%) were diagnosed electrophysiologically as definite CIDP. **Conclusion:** CIDP affects males mostly in the 3rd to 5th decades. Typical CIDP was most common. Weakness in both proximal and distal muscle groups along with paresthesia were the most common symptoms. Few patients had distal wasting. Diabetes mellitus was associated disease in many cases. *[Journal of National Institute of Neurosciences Bangladesh, January 2025;11(1):34-40]*

Keywords: Chronic inflammatory demyelinating polyradiculoneuropathy; CIDP; electrophysiology, weakness; paresthesia

Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is the most common chronic immune-mediated inflammatory polyneuropathy, with a heterogeneous clinical manifestation[1-3]. It is characterized by progressive or relapsing motor or sensory symptoms. CIDP classically presents with progressive, relatively symmetric proximal and distal muscles weakness, generalized areflexia, and large fiber sensory loss[4]. According to the definition of the European Federation of Neurological Societies and the Peripheral Nerve Society (EFNS/PNS), CIDP is progressive or relapsing disease over 2 months, has electrophysiological or pathological evidence of peripheral nerve demyelination, and responds to immunosuppressive or immune-modulating therapies[5,6]. CIDP is considered an orphan disease with different prevalence rates in different geographical regions[7-9]. One

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2019 meta-analysis estimated the incidence rate to be 0.33 per 100,000 and the prevalence rate to be 2.81 per 100,000[10]. In that study male patients outnumbered female patients by 2:1, and CIDP was more prevalent in advanced age[8].

In 2005, the Joint Task Force of the European Federation of Neurological Societies and the Peripheral Nerve Society (EFNS/PNS) classified CIDP into clinical subtypes. In this guideline (updated in 2010)[11], CIDP is categorized as 'typical CIDP', and 'atypical CIDP'. Typical CIDP is a symmetrical polyneuropathy affecting proximal and distal muscles equally, whereas atypical CIDP includes distal acquired demyelinating symmetric (DADS), and multifocal acquired demyelinating sensory and motor neuropathy (MADSAM/asymmetric CIDP, or Lewis-Sumner syndrome [LSS]) and pure motor or sensory CIDP. DADS is a symmetrical length-dependent sensory or sensorimotor neuropathy, often associated with an IgM para protein and markedly increased distal motor latencies. European Academy of Neurology/ Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force-Second revision[12], divided CIDP into typical CIDP and CIDP variants. The previous term atypical CIDP was replaced by CIDP variants because these are well-characterized entities like multifocal, focal, distal, motor, or sensory CIDP. The levels of electrophysiological diagnostic certainty were reduced from three, like definite, probable, and possible CIDP to only two, like CIDP and possible CIDP.

Basic serum laboratory studies should be performed to exclude alternative or confounding diagnoses, including hemoglobin A1c, complete blood cell count, electrolytes, liver, renal, and thyroid function studies, and vitamin B12 and methylmalonic acid levels. All patients with possible CIDP must be screened for a monoclonal gammopathy with serum and urine electrophoresis, immunofixation, and free light chains. CSF analysis is usually not necessary, but, in certain cases of suspected CIDP, it may be helpful. When CSF is collected, albumino-cytological dissociation (elevated protein, normal leukocyte counts) is anticipated.

CIDP can lead to severe disability if left untreated[13-15]. Immunoglobulin and plasma exchange are considered effective first-line treatment for CIDP[16-19]. However, up to 21.0% patients with CIDP are refractory to these immune therapies and have severe disabilities. The aim of this study was to evaluate the clinical and electrophysiological aspects of CIDP in adult.

Methodology

Study Settings and Population: This cross-sectional study was carried out in the Department of Neurophysiology of National Institute of Neurosciences and Hospital, Bangladesh from July 2020 to June 2022 for a period of two years. Clinically suspected cases of CIDP were referred to the Neurophysiology lab for nerve conduction study. Patient of 18- 70 years of aged fulfilling EFNS /PNS diagnostic criteria for CIDP were included in this study. Detail history was taken and thorough examinations were done in every patient.

Study Procedure: For all the enrolled patient, clinical parameters including age, sex, antecedent events, course of disease, sensory symptoms, associated diseases like diabetes mellitus (DM), weakness and wasting of muscles, sensory disturbances, reflexes, cranial nerve deficits, autonomic dysfunction and treatment modality were collected.. Muscle weakness of 4 limbs were evaluated by the Medical Research Council (MRC) scale and the final MRC sum score {the total range 0 (complete paralysis) to 60 (normal strength)} was assessed in every patient as well as CSF study and neurophysiological studies were done in all patients.

Electrophysiology: Motor nerve conduction studies and F wave studies were performed in the median, ulnar, tibial and peroneal nerves using conventional procedures. Antidromic sensory nerve conduction studies were conducted in the median, ulnar and sural nerves. According to the electro diagnostic criteria for demyelination proposed by EFNS/PNS, 2010 the presence of demyelinating conduction abnormalities of the median and ulnar nerves was determined in the two segments; distal nerve segments (distal to the wrist) and intermediate nerve segments (wrist to elbow). The terminal latency index was used to compare the distal segment with the intermediate segment, and was calculated using the formula: Distal conduction distance (mm)= forearm conduction velocity (m/s) /distal latency (ms). In sensory nerve conduction studies, we focused on the involvement patterns of the median, ulnar and sural sensory nerve responses. The pattern of 'abnormal median-normal sural sensory responses' suggests demyelination predominant in the distal nerve terminals, and is specifically seen in patients with CIDP or demyelinating Guillain-Barre syndrome. Statistical analyses were performed using the SPSS version 22.0 for Windows program.

Statistical Analysis: Statistical analysis was performed by Windows based software named as Statistical Package for Social Science (SPSS), versions 22.0 (IBM

SPSS Statistics for Windows, Version 22.0. Armonk, NY: IBM Corp.). Continuous data were expressed as mean, standard deviation, minimum and maximum. Categorical data were summarized in terms of frequency counts and percentages. Chi-square test was used for comparison of categorical variables and Student t test was applied for continuous variables. Every efforts were made to obtain missing data. A two-sided P value of less than 0.05 was considered to indicate statistical significance. Differences between case and control were tested.

Ethical Consideration: All procedures of the present study were carried out in accordance with the principles for human investigations (i.e., Helsinki Declaration 2013) and also with the ethical guidelines of the Institutional research ethics. Formal ethics approval was granted by the local ethics committee. Participants in the study were informed about the procedure and purpose of the study and confidentiality of information provided. All participants consented willingly to be a part of the study during the data collection periods. All data were collected anonymously and were analyzed using the coding system.

Results

A total of 50 patients fulfilling EFNS diagnostic criteria for CIDP, 2010 were enrolled in this study. The mean age was 43.82 ± 13.43 yrs. Most of the patients were male. The age of onset ranged from 20-70 years and mean age (Mean ± SD) was 43.82 ± 13.43 years. Males (72%) were more affected. DM was the most common (36%) associated disease (Table 1).

Table 1: Distribution of the study subjects (n=50)

Variables	Frequency	Percent
Age Group		
• 20 to 30 Years	10	20.0
• 31 to 40 Years	12	24.0
• 41 to 50 Years	14	28.0
• 51 to 60 Years	9	18.0
• 61 to 70 Years	5	10.0
Mean ± SD	43.82 ± 13.43	
Gender		
• Male	36	72.0
• Female	14	28.0
Residence		
Urban	15	30.0
• Rural	35	70.0
Co-morbidity of the study subjects		
Diabetes mellitus	18	36.0

Symptoms and Signs at Presentation: Most patients (96.0%) presented with sensory motor polyneuropathy and two patients(4.0%) had a sensory neuropathy. Weakness of limbs was present in forty-eight patients (96%) which was symmetrical (table-2) and weakness was ranged from mild weakness of intrinsic muscles of hands and feet to severe weakness becoming bed-bound (table 3).Wasting was present in 30% of patient with a distal predominance .(table-2).). Sensory impairment in the form of paresthesia was present in 100% of patients. Tingling and numbness was the most (68.0%) common

Table 2: Clinical Features and Examination Findings of the Disease of the Study Subjects (n=50)

Variables	Frequency	Percent
Symptoms of the Disease		
Weakness		
• All limb	48	96.0
• No weakness	2	4.0
Wasting	15	30.0
• Upper limb-distal	6	12.0
• All limbs-distal	9	18.0
Sensory symptoms(paresthesia)		
• Tingling	3	6.0
• Numbness	5	10
Tingling and numbness	34	68.0
Tingling plus numbness plus burning sensation	8	16.0
Temporal course		
• Progressive slowly	39	78.0
• Progressive rapidly	7	14.0
• Relapsing	4	8.0
Examination Findings		
Pain sense		
• Impaired	44	88.0
• Intact	6	12.0
Touch sense		
• Impaired	44	88.0
• Intact	6	12.0
Position sense		
• Intact	34	68.0
• Impaired	16	32.0
Vibration sense		
• Intact	29	58.0
• Impaired	21	42.0
Romberg's sign (Present)	14	28.0
Gait		
• Impaired	38	76.0
• Normal	12	24.0
Duration of disease (Mean ± SD)	17.41 ± 25.25 months	

symptoms. About 88% patients had clinical evidence of sensory impairment. Among them 88.0% patients had impairment of pain and touch, 32.0% had impairment of position and vibration sense (Table 2).

Table 3: Power of Muscles of the Study Subjects (n=50)

Power of Muscles	Mean ± SD	Min–max (MRC grade)
Upper arm abductors	4.20 ± 0.93	0- 5.00
Elbow flexors	4.20 ± 0.93	0 - 5.00
Wrist extensors	3.52 ± 1.15	0 - 5.00
Hip flexors	4.00 ± 0.93	0 - 5.00
Knee extensors	4.00 ± 0.93	0 - 5.00
Foot dorsal flexors	3.28 ± 1.13	0 - 5.00

Clinical Course: The duration of illness was 17.41 ± 25.25 months. On the basis of history, the patients were classified into two major subgroups, relapsing and non-relapsing. The non-relapsing group consisted of 46 patients(92.0%) in whom there were no relapses who had either sub-acute onset and slowly progressive (78.0%) or sub-acute onset and rapidly progressive (14%). The relapsing group consisted of 4 patients (8%) in whom there was a history of definite relapse and remission. In relapsing group earlier age group (20-30 years' age group) was more predominant (Figure A, B, C).

Electrophysiology: Nerve conduction study revealed prolonged distal motor latency in median, ulnar, tibial and peroneal nerves {median nerve (mean±SD) 8.81±6.25ms, ulnar - 6.27±3.63 ms, tibial 7.19±5.33ms, peroneal. 7.99±6.65ms}, normal to smaller distally evoked compound muscle action potential, normal to reduced conduction velocity in all studied nerves. In sensory nerve conduction study, normal to prolonged sensory latency in studied nerves, the pattern of abnormal median-normal sural sensory response was present in 8% of patients in typical CIDP (Table 5).

Table 5: Nerve Conduction Study of the Subjects (n=50)

Nerve Conduction Study	Mean ± SD	Min - max
Motor NCS		
Motor median		
• Distal latency (ms)	8.81 ± 6.25	3.26 - 33.50
• F- wave latency (ms)	34.07 ± 28.17	0.00 - 100.00
• Conduction velocity(m/s)	32.54 ± 13.11	0.00 - 56.00
• CMAP amplitude(mV)	7.13 ± 6.37	0.53 - 35.00
Ulnar motor		
• Distal latency(ms)	6.27 ± 3.63	2.08 - 17.00
• F-wave latency(ms)	30.18 ± 24.51	0.00 - 70.45
• Conduction velocity(m/s)	32.32 ± 14.59	0.00 - 67.80
• CMAP amplitude(mV)	4.88 ± 3.31	0.00 - 12.50
Tibial motor		
• Distal latency(ms)	7.19 ± 5.33	0.00 - 21.50
• F-wave latency(ms)	30.05 ± 32.71	0.00 - 100.00
• Conduction velocity(m/s)	25.09 ± 12.42	0.00 - 50.00
• CMAP amplitudes(mV)	4.19 ± 4.72	0.08 - 18.00
Peroneal Nerve		
• Distal latency (ms)	7.99 ± 6.65	0.00 - 33.00
• Conduction velocity(m/s)	22.33 ± 9.97	0.00 - 40.00
• CMAP amplitude	1.16 ± 1.35	0.00 - 4.60
Sensory NCS		
Median Nerve		
• _DL	3.55 ± 0.66	2.50 - 4.56
• SNAP	16.20 ± 16.76	3.00 - 60.00
Ulnar nerve		
• DL	3.57 ± 1.83	2.38 - 7.20
• SNAP	14.99 ± 13.06	2.00 - 38.70
Sural Nerve		
• _DL	3.17 ± 0.91	2.19 - 4.14
• _SNAP	6.43 ± 3.58	3.00 - 11.00
Abnormal median –normal sural	4	8%

CMAP: Compound Muscle Action Potential, SNAP: Sensory Nerve Action Potential, DL-distal latency.

Most patients presented with typical CIDP (90.0%). Among the atypical cases 4.0% patients were MADSAM, 4.0% sensory CIDP and 2.0% DADS (Table 6).

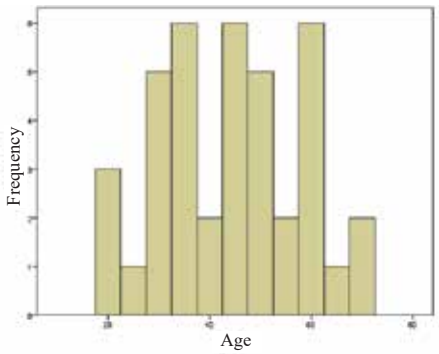

Figure A: Progressive slowly

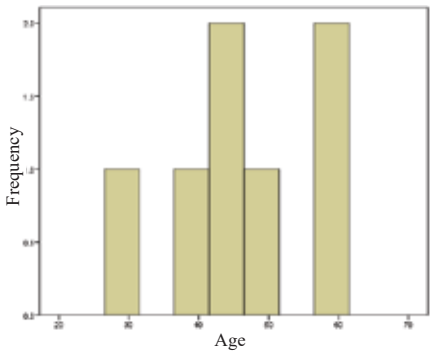

Fig. B: Progressive rapidly

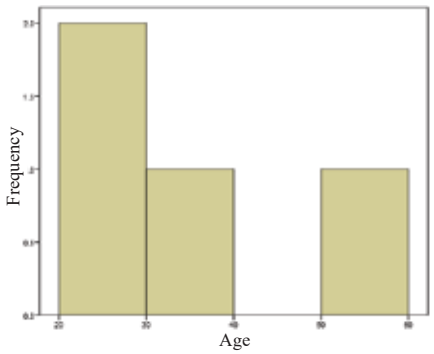

Figure C: Relapse

Table 6: Subtypes of CIDP

Variants of CIDP	Frequency	Percent
Typical CIDP	45	90.0
Sensory CIDP	2	4.0
MADSAM	2	4.0
DADS	1	2.0

MADSAM: multifocal acquired demyelinating sensory and motor neuropathy, DADS: distal acquired demyelinating symmetric neuropathy. CIDP: chronic inflammatory demyelinating polyneuropathy.

According to European Academy of Neurology/ Peripheral Nerve Society (EAN/PNS) 2021 electrodiagnostic criteria, 92.0% patients had CIDP and rest of patients were possible CIDP (Table 7).

Table 7: EAN/PNS Electro Diagnostic Criteria 2021

EAN/PNS electrodiagnostic criteria	Frequency	Percent
CIDP	46	92.0
Possible CIDP	4	8.0

EAN/PNS: European Academy of Neurological and the Peripheral Nerve Society.

CSF Study: CSF examination was done in every patient. CSF protein levels were 35.00 to 545.00mg/dl (mean 139.68 ± 87.27). Albumino-cytological dissociation was reported in 95.0% patients (Table 8).

Table 8: CSF study of the study subjects (n=50)

CSF study	Mean ± SD	Min - max
Protein	139.68 ± 87.27	35.00 - 545.00
Cell_count	2.52 ± 1.70	2.00 - 10.00
Albumino-cytological dissociation	47(frequency)	95(Percentage)

CSF: cerebrospinal fluid.

Discussion

In this study, 50 patients, with the clinical diagnosis of CIDP over a period of 2 years were recruited and we reviewed the clinical, electrophysiological and laboratory findings of the patients who fulfilled the criteria for diagnosis of CIDP according to EFNS/PNS 2010 modified by European Academy of Neurology/Peripheral Nerve Society (EAN/PNS 2021). They had a predominantly symmetric sensorimotor neuropathy with generalized areflexia/hyporeflexia and there were electrophysiological features of peripheral nerve demyelination and mostly an elevated CSF protein. This study showed a male predominance (72.0%) with sex ratio being 2.6:1 which is consistent with other studies of CIDP[8]. In this study the average age of onset was 43.82 ± 13.43 years. McCombie et al[20] found mean age of onset was 35.4 SD 20.3 years in their study. The average age of onset was found to be 23 years and 26 years in the study by Austin et al[21] and Thomas et al[22]. In this study the majority of cases occurred in 30 to 50 years of age group but there was a significantly earlier age of onset (20-30 year) in patients of relapsing condition observed that was consistent with other study[20]. The study by McCombie et al[20] found an increased number of relapsing varieties of CIDP in 10 to 30 years of age group. In this study 36.0% patients had associated with Diabetes mellitus. Diabetes mellitus (DM) was associated with CIDP in 9.0% to 26.0% of CIDP patients in different study[21-25].

We have classified our cases into relapsing and non-relapsing groups using criteria similar to those described by others[26-30]. Cases with subacute onset and slowly or rapidly progressive and monophasic course have been included in the non-relapsing group. Patients who had definite relapses with or without complete recovery are included in the relapsing group. 46(92.0%) patients had a non- relapsing/gradually progressive course and 4(8.0%) patients had relapsing course. This finding is somewhat contradictory to the findings in a study where a relapsing course was found in 30.0% of cases, a chronic and progressive course in 60.0%, and a monophasic with full recovery in 10.0% cases[31].

The most common initial symptoms were tingling and numbness which was distally predominant. Weakness of proximal and distal muscles and paresthesia were the most common symptoms. Weakness, which was symmetrical and ranged from mild to severe, was present in 96.0% of the patients. Wasting was present in 30.0% of patients which was distally predominant. Paresthesia was present in all patients. 88.0% patients had clinical evidence of sensory impairment. Study by McCombie et al[20] found weakness to be present in 94.0% patient and paresthesia in 64.0% of the patients[20]. Most patients presented with typical CIDP (90.0%). Among the atypical cases 4.0% patients were MADSAM, 4.0% sensory CIDP and 2.0% DADS. The Italian CIDP Database study group analyzed data from 460 patients with CIDP, and found that 82.0% had typical CIDP and the remaining 18.0% had atypical CIDP which included DADS (7.0%), MADSAM (4.0%), pure motor (4.0%) and pure sensory CIDP (3.5%)[32]. In a Japanese study[33], 100 consecutive patients with CIDP were classified as having typical CIDP (60.0%), MADSAM (34.0%), DADS (5.0%) or

pure sensory CIDP (1.0%).

Electrophysiology in patients with typical CIDP showed a longer distal motor latency, smaller distally evoked compound muscle action potential, reduced conduction velocity. In sensory nerve conduction study, the pattern of abnormal median-normal sural sensory response was present in 8.0% of patients in typical CIDP which differs from the study by Kuwabara et al[33] where they found an abnormal median-normal sural sensory response in 53.0% of typical CIDP and 13.0% in atypical CIDP.

CSF examination was done in every patient. CSF protein levels were 35.00-545.00mg/dl (mean 139.68±87.27). This finding matches well with the observation of McCombie et al[20] who found CSF protein between 0.23 to 5.5 g/l. In our study albuminocytologic dissociation (CSF protein >45 mg/dl and the CSF white cell count is normal called albuminocytologic dissociation) in CSF was present in 94.0% of the patients. In different studies[33-36], albuminocytologic dissociation in CSF was present in more than 80.0 % of patients with CIDP.

Conclusion

CIDP affects males mostly in the 3rd to 5th decades. Weakness in both proximal and distal muscle groups along with paresthesia were the most common symptoms. Few patients had distal wasting. DM was associated disease in nearly one-third of the patients. Most of patients fulfilled the diagnostic criteria of typical CIDP. The course of disease was progressive in most of patients with few patients had h/o relapsing in course. The majority of patients were diagnosed electrophysiologically as definite CIDP.

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Conflict of interest: Authors declared no conflict of interest.

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Contribution to authors: Sumi MSN, Mamun AA, Hussain ME, Habib MA conceived and designed the study, analyzed the data, interpreted the results, and wrote up the draft manuscript. Habib MA, Khan AFMAM, Shaikh MMI, Islam MN involved in the manuscript review and editing. Alam MB, Mohammad QD, Chowdhury RN conceived and manuscript writing. All authors read and approved the final manuscript.

Data Availability
Any inquiries regarding supporting data availability of this study should be directed to the corresponding author and are available from the corresponding author on reasonable request.

Ethics Approval and Consent to Participate
Ethical approval for the study was obtained from the Institutional Review Board. As this was a prospective study the written informed consent was obtained from all study participants. All methods were performed in accordance with the relevant guidelines and regulations.

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