Clinical Characteristics and Treatment Response of Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Background: *The incidence, prevalence, clinical phenotypes, and treatment response of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) are varying in the world literature. There have been no epidemiologic studies of CIDP in Thai adult patients.*

Objective: *To determine clinical characteristics, phenotypes, electrophysiological tests, and treatment response of CIDP in Thai adult patients and to find factors associated with disease outcome after treatment.*

Material and Method: *Retrospective chart review of Prasat Neurological Institute patients diagnosed of CIDP between January 1, 2008 and December 31, 2014.*

Results: *Sixty-three CIDP patients were identified. Patients were slightly male predominant (1.3:1), age at onset was 47.7 years, disease duration prior to first evaluation was 5.0 months, follow-up duration was 26.8 months, and 19% of patients had diabetes. Clinical phenotypes were classic CIDP (76.2%), 19% DADS, and 4.8% MADSAM. Fifteen point nine percent presented as AIDP and 12.7% as SIDP. Symmetrical, sensorimotor polyneuropathy with hyporeflexia were the common presentation. Autonomic symptoms, respiratory failure, bulbar involvement, ophthalmoparesis, ptosis, and muscle atrophy were rarely presented. The treatment response was generally favorable. Patients in disease relapsing group had shorter disease onset (2 vs. 6 months) and 40% had disease duration less than four weeks.*

Conclusion: *Clinical characteristics, phenotypes, electrophysiological findings, and treatment response of CIPD in Thai patients were not different from previously published studies in western and oriental populations. Mode of disease onset may predict a response to immunosuppressive treatment in CIDP patients.*

Keywords: *Chronic inflammatory demyelinating polyradiculoneuropathy, CIDP, Thai patients, Treatment response*

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a symmetric, motor predominant, proximal, and distal demyelinating peripheral neuropathy. CIDP is caused by an inflammatory or immune response against myelin proteins in the peripheral nervous system affecting spinal nerve roots, plexus and peripheral nerves. The clinical course of CIDP lasts more than eight weeks and can be classified into three types, monophasic, slowly progressive, and relapsing course[1,2]. The prevalence and incidence of CIDP have greatly varied among geographic location and countries. The prevalence of CIDP ranged from 1.9 to 7.7 per 100,000 persons[3-6] and the incidence ranged from 0.15 to 0.48 per 100,000 person-years[6-8]. The prevalence was greater in males than females. The mean age of onset was 47.6 years[4]. Several variants of CIDP have been described based on the distribution of symptoms and signs[9]. The main classification proposed by Saperstein et al included: 1) classic CIDP, 2) distal acquired demyelinating symmetric neuropathy (DADS), 3) multifocal motor neuropathy (MMN), and 4) multifocal acquired demyelinating sensory and motor neuropathy (MADSAM)[10]. The response to treatment and long-term prognosis of CIDP highly depend on age at the onset and clinical phenotypes[9]. Recognition of CIDP is important because CIDP is a treatable disease and many patients respond to immunosuppressive or immunomodulation therapies[11-13]. Obtaining reliable information regarding the incidence, prevalence, clinical phenotypes, and treatment response of CIDP are necessary to realize the current situation in Thailand and to plan for the health care needs and costs. Currently, the epidemiologic studies of CIDP in Thailand have not yet been reported.

The primary goal of the present study was to determine the clinical characteristics, clinical phenotypes, electrophysiological findings, and treatment response of CIDP in Thai adult patients in a single

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center. The secondary goal was to compare CIDP patients who got a remission after immunosuppressive treatment versus CIDP patients who relapsed after treatment in order to find factors associated with disease outcome after treatment. The result from the present study would be useful for the prediction of disease outcome after treatment.

Material and Method

After the Institutional Review Board (IRB) approval, the disease diagnoses registries were searched for the potential diagnosis of CIDP or demyelinating neuropathy dating between January 1, 2008 and December 31, 2014. CIDP patients were diagnosed using criteria proposed in 2010 by the European Federation of Neurological Societies/Peripheral Nerve Society in 2010 (EFNS/PNS guideline). This diagnostic criterion consists of clinical diagnostic criteria, electrodiagnostic criteria, and supportive criteria[12].

For diagnosis of CIDP, patients have to meet the clinical diagnostic criteria and electrodiagnostic criteria with or without supportive criteria as described in the guideline. In the EFNS/PNS guideline, patients were categorized into definite, probable, and possible CIDP, based on the combination of clinical diagnostic criteria, electrodiagnostic criteria, and supportive criteria. All CIDP patients including definite, probable, and possible CIDP were included in the present study.

The authors did not exclude CIDP patients with diabetes and multifocal motor neuropathy patients from the present study. Because the authors aimed to evaluating clinical characteristics of all CIDP phenotypes and associated disease in Thai adult patients. Identifying CIDP in diabetic patients is difficult because of diabetic polyneuropathy (DPN) can demonstrated demyelinating features on nerve conduction studies as well as raised cerebrospinal fluid (CSF) protein. To date, there is no criterion to confirm diagnosis of CIDP in diabetes patients. The authors included diabetes patients to the present study if patients had progressive symmetric, painless, sensorimotor polyneuropathy. If diabetes patients had one of the following features, including painful, asymmetric, distal sensory predominant polyneuropathy, they will be excluded from the present study.

Once the CIDP patients were identified, the medical records were reviewed to assess demographic features, clinical manifestation, clinical phenotypes proposed by Saperstein et al[10], disease duration prior to first evaluation, follow-up duration, clinical course, laboratory results including electrodiagnostic studies and CSF profiles, immunosuppressive treatment, and treatment response. Disease severity and disability were assessed and graded using modified ranking scale (mRS). The authors classified patients into 3 subgroups, mild disability with functional independence (mRS score 0 to 2), moderate disability with functional partial dependence (mRS score 3) and severe disability with functional dependence (mRS score 4 to 5).

To find factors associated with disease outcome after treatment, the authors classified patients into two groups: 1) patients with disease remission or stable after immunosuppressive treatment and 2) patients with disease relapsing after immunosuppressive treatment. The authors compared patients in both groups to find a predictor for the treatment response in CIDP. Disease remission means asymptomatic or stable disease activity in the patients who had been discontinued treatment more than 1 year. Stable disease means stable or improved disease activity in the patients who received immunosuppressive treatment more than 3 months and need to continue the treatment. Disease relapsing means relapsed disease in patients who had been off treatment or remained on treatment[14].

Statistical analysis

Descriptive summaries were presented as frequencies and percentages for categorical variables and median/mean and ranges for continuous variables. Comparisons between CIDP patients with remission after immunosuppressive treatment versus CIDP patients with relapsing disease after treatment were performed using Fisher's exact test or Wilcoxon rank sum test, as appropriate. All of the tests were two sided, and p-value less than 0.05 were considered as statistical significance.

Results

Demographic characteristics

From the disease registry between January 1, 2008 and December 31, 2014 at Prasat Neurological Institute, 63 CIDP patients were identified using diagnostic criteria. The demographic and clinical characteristics have been shown in Table 1.

Characteristics of polyneuropathy symptoms

The characteristics of polyneuropathy symptoms have been presented in Table 1. All patients were categorized into definite CIDP. Out of 63 patients, 15.9% presented at first evaluation as acute onset

Table 1. The demographic and clinical characteristics of CIDP patients

	Total n = 63	Remission n = 43	Relapsing n = 20	*p*-value	OR	95% CI
Sex (male:female)	1.3:1	1.9:1	1:1.5	0.061	0.36	0.12 to 1.07
Age at onset (years), mean (SD)	47.7 (17.1)	47.4 (17.6)	48.5 (16.2)	0.806	1.00	0.97 to 1.04
Age at evaluation (years), mean (SD)	48.5 (17.0)	48.0 (17.7)	49.4 (15.9)	0.765	1.01	0.97 to 1.04
Duration prior to 1st evaluation (months), median (IQR 25, 75)	5.0 (2.0, 9.6)	6.0 (2.9, 10.4)	2.0 (0.5, 5.9)	0.010	1.01	0.98 to 1.04
Less than 4 weeks (%)	15.9	4.7	40.0	0.001	-	-
Between 4 to 8 weeks (%)	12.7	11.6	15.0		0.15	0.02 to 1.24
More than 8 weeks (%)	71.4	83.7	45.0		0.06	0.01 to 0.35
Follow-up duration (months), median (IQR 25, 75)	26.8 (8.6, 39.0)	16.8 (6.1, 35.9)	37.6 (21.2, 86.9)	0.040	1.03	1.01 to 1.05
Underlying disease (%)						
No underlying disease	69.8	60.5	90.0	0.017	5.89	1.21 to 28.7
Diabetes	19.0	23.3	10.0	0.309	0.37	0.07 to 1.86
HIV	4.8	7.0	0.0	0.545	0.00	0.00 to 0.00
Others	7.9	11.6	0.0	0.169	0.00	0.00 to 0.00
CIDP phenotypes (%)						
Classic CIDP	76.2	76.7	75.0	0.406	-	-
DADS	19.0	20.9	15.0		0.73	0.17 to 3.10
MADSAM	4.8	2.3	10.0		4.40	0.37 to 52.38
Clinical manifestation (%)						
Sensorimotor	84.1	83.7	85.0	0.317	-	-
Pure motor	4.8	2.3	10.0		0.24	0.02 to 2.79
Pure sensory	11.1	14.0	5.0		0.08	0.00 to 2.05
Autonomic symptoms	0.0	0.0	0.0	NA	-	-
Respiratory failure	1.6	2.3	0.0	1.000	0.00	0.00 to 0.00
Ophthalmoparesis	3.2	2.3	5.0	0.538	2.21	0.13 to 37.25
Ptosis	1.6	0.0	5.0	0.317	0.00	0.00 to 0.00
Bulbar involvement	3.2	2.3	5.0	0.538	2.21	0.13 to 37.25
Symmetrical	95.2	97.7	90.0	0.230	0.21	0.02 to 2.52
Hyporeflexia or areflexia	93.7	93.0	95.0	1.000	1.43	0.14 to 14.62
Muscle atrophy	15.9	18.6	10.0	0.481	0.47	0.09 to 2.46
Distribution of motor weakness (%)						
Proximal greater or equal to distal	61.9	58.1	70.0	0.521	-	-
Distal greater than proximal	27.0	27.9	25.0		2.50	0.24 to 26.48
No motor weakness	11.1	14.0	5.0		3.36	0.37 to 30.81
Sensory symptom (%)						
Negative sensory symptom	95.2	97.7	90.0	0.234	0.21	0.02 to 2.52
Pain	15.9	11.6	25.0	0.266	2.53	0.64 to 10.03
Definite CIDP by EFNS/PNS criteria (%)	100.0	100.0	100.0	NA	-	-

CIDP = chronic inflammatory demyelinating polyradiculoneuropathy; DADS = distal acquired demyelinating symmetric; MADSAM = multifocal acquired demyelinating sensory and motor; EFNS/PNS = European Federation of Neurological Societies/ Peripheral Nerve Society; NA = not applicable

CIDP mimicking acute inflammatory demyelinating polyradiculoneuropathy (AIDP) with disease onset less than four weeks. A further 12.7% presented as subacute inflammatory demyelinating polyradiculoneuropathy (SIDP) with disease onset between 4 to 8 weeks and 71.4% as CIDP with disease onset more than eight weeks. The clinical phenotypes were then analyzed. The majority of patients had classic CIDP (76.2%). In contrast, 19% had DADS and only 4.8% had MADSAM. The analysis of the pattern of polyneuropathy revealed that the majority of patients had symmetrical, sensorimotor polyneuropathy with hyporeflexia or areflexia. Patients presented with pure sensory and pure motor neuropathy were found in approximately 11.1% and 4.8% of all patients, respectively. Autonomic symptoms (0%), respiratory failure (1.6%), bulbar involvement (3.2%), ophthalmoparesis (3.2%), ptosis (1.6%), and muscle atrophy (15.9%) were uncommon manifestations in the present study. The analysis of the distribution of motor weakness showed that the majority of patients (61.9%) had proximal muscle weakness and the weakness was either predominant over or equal to distal muscle weakness. Predominant distal muscle weakness could be found in only 27% of

Table 2. Electrodiagnostic studies and CSF profiles of CIDP patients

	Total	Remission	Relapsing	*p*-value	OR	95% CI
EMG criteria (EFNS/PNS criteria) (%)	n = 63	n = 43	n = 20			
Definite	93.7	95.3	90.0	0.586	2.28	0.30 to 17.46
Possible	6.3	4.7	10.0			
Nerve conduction study abnormality (%)	n = 63	n = 43	n = 20			
Conduction block	33.3	30.2	40.0	0.444	1.54	0.51 to 4.65
Slow conduction velocity	79.4	79.1	80.0	1.000	1.06	0.28 to 3.96
Prolonged F-wave	79.4	79.1	80.0	1.000	1.06	0.28 to 3.96
Prolonged distal latency	76.2	74.4	80.0	0.908	1.38	0.38 to 5.01
Terminal latency index (TLI) less than 26	28.6	27.9	30.0	0.864	1.11	0.35 to 3.55
Cerebrospinal fluid (CSF) examination	n = 59	n = 40	n = 19			
Protein (mg/dL), mean (SD)	146.4 (98.0)	160.3 (107.4)	117.0 (67.8)	0.065	1.00	0.99 to 1.00
Glucose (mg/dL), median (IQR 25, 75)	67.0 (55, 74)	68.0 (57, 79)	57.0 (52, 69)	0.050	0.95	0.91 to 1.00
Leukocyte (cell/μL), median (IQR 25, 75)	0 (0, 2)	0 (0, 2)	1 (0, 3)	0.451	0.98	0.86 to 1.12
Mononuclear cell (%), median (IQR 25, 75)	100 (100, 100)	100 (100, 100)	100 (99.5, 100)	0.559	1.09	0.67 to 1.77
Elevated CSF protein with leukocyte count less than 10 cell/μL (%)	84.7	85.0	84.2	1.000	0.94	0.21 to 4.25

EMG = electromyography

all patients. The sensory abnormalities were analyzed and the results showed most patients had negative sensory symptom or decreased sensation (95.2%) and pain was present in 15.9% of the patients.

Electrodiagnostic studies and CSF profiles

The electrodiagnostic studies and CSF profiles were presented in Table 2.

Treatment outcome after immunosuppressive therapy

The patients in the present study received variable regimens of treatment (Table 3). Most patients were treated with prednisolone (33.3%) or prednisolone and azathioprine (19.0%). Others were intravenous immunoglobulin (12.7%), monthly intravenous pulse methylprednisolone (9.5%), plasmapheresis (1.6%), or combinations of the above mentioned (23%). The follow-up periods of each patient were also varied. The median follow-up duration was 26.8 months. The median follow-up duration before first recurrence was 13.7 months.

The severity of the disease at the onset (before treatment) and at the last visit (after treatment) were analyzed and were presented in Table 3. Out of 63 patients, 12.7% had mild disability at the onset (mRS = 0 to 2), 36.5% had moderate disability (mRS = 3), and 50.8% had severe disability (mRS = 4 to 5). After they received immunosuppressive treatment, many patients had good response to treatment. Out of 63 patients, 71.4% had mild disability (mRS = 0 to 2), 20.6% had moderate disability (mRS = 3), and 7.9% had severe disability (mRS = 4 to 5) at the last visit.

Prognostic factors after immunosuppressive therapy in CIDP patients

Out of 63 patients, 43 (68.3%) had disease remission and 20 (31.7%) had disease relapse. The median follow-up duration before first recurrent was 13.7 months (Table 3). To study prognostic factors in CIDP, the authors classified patients into two groups: 1) patients with disease remission or stable after immunosuppressive treatment and 2) patients with disease relapsing after immunosuppressive treatment. The authors compared patients in both groups to find a predictor for the treatment response in CIDP. The disease relapsing group had shorter disease duration prior to first evaluation (2 vs. 6 months, $p = 0.010$). Forty percent of patients in relapsing group presented as acute onset CIDP mimicking AIDP with disease duration prior to diagnosis less than four weeks. These patients received treatment as AIDP including intravenous immunoglobulin (IVIg) and plasmapheresis. Patients in relapsing group were trend to have more severe disability than patients with disease remission but did not reach statistical significance (70.0 vs. 41.8%, $p = 0.119$). Other factors regarding clinical features, electrodiagnostic findings, CSF profiles, and treatment regimens were not different between both groups.

Discussion

The prevalence, incidence and clinical phenotypes of CIDP are varied among publications. These could be related to several factors, including genetic predisposition, using different clinical spectrum, and different diagnostic criteria[9]. The

Table 3. Treatment and responsive to treatment of CIDP patients

	Total n = 63	Remission n = 43	Relapsing n = 20	*p*-value	OR	95% CI
Treatment (%)						
Prednisolone	33.3	37.2	25.0	0.155	-	-
Prednisolone and azathioprine	19.0	23.2	10.0		2.45	0.71 to 8.43
Others*	47.7	39.5	65.0		0.64	0.10 to 3.95
mRS at onset (%)						
Mild (0 to 2)	12.7	14.0	10.0	0.119	-	-
Moderate (3)	36.5	44.2	20.0		0.63	0.09 to 4.35
Severe (4 to 6)	50.8	41.8	70.0		2.33	0.41 to 13.38
mRS at last follow-up (%)						
Mild (0 to 2)	71.4	69.8	75.0	1.000	-	-
Moderate (3)	20.6	20.9	20.0		0.89	0.24 to 3.36
Severe (4 to 6)	7.9	9.3	5.0		0.50	0.05 to 4.88
Number of recurrent, median (IQR 25, 75)	0 (0, 1)	0 (0, 0)	1 (1, 2)	<0.001	NA	-
Follow-up duration before 1st recurrence (months), median (IQR 25, 75)	13.7 (3.9, 32.1)	16.6 (6.0, 35.3)	6.2 (1.6, 23.0)	0.037	0.99	0.97 to 1.01

mRS = modified ranking scale

* Other treatments included intravenous immunoglobulin (12.7%), monthly intravenous pulse methylprednisolone (9.5%), plasmapheresis (1.6%), combinations of the previous mentioned (23%), and no treatment (0.9%)

diagnostic criteria for CIDP is mainly based on clinical characteristics and demyelinating features from nerve conduction studies, which are supported by nerve biopsy, albumino-cytological dissociation in CSF, evidence of gadolinium enhancement, and/or hypertrophy of nerve roots in magnetic resonance imaging and clinical improvement following immuno-modulatory treatment[12]. Now, at least 15 diagnostic criteria for CIDP have been proposed. In the present study, the authors used the diagnostic criteria for CIDP, proposed in the EFNS/PNS guideline because they included other variants in the criteria and demonstrated a high sensitivity and specificity for CIDP diagnosis[15]. The present study demonstrated clinical and electrodiagnostic features of CIDP in Thai patients. The results showed that the demographic, characteristics of polyneuropathy, electrodiagnostic features, and CSF profiles were not different from previously published studies in western and in oriental populations[4,16]. The clinical phenotypes of patients in the present study were mostly similar to previously published studies[16-21]. However, there were some differences including: 1) the MMN phenotype was not found in the present study and 2) DADS phenotype was higher than the study from Malaysia[16]. These findings may reflect the difference phenotypes of CIDP between racial and ethnic groups or may be a result of confounding and information bias in the present study. Future studies are required.

In the present study of 63 patients, 12 patients had diabetes (19%). All CIDP with diabetes patients had symmetric, painless, and sensorimotor poly-neuropathy in all extremities with progressive motor weakness. These patients had electrodiagnostic features and CSF profiles compatible with those proposed by the EFNS/PNS guideline in 2010. Eleven of 12 CIDP with diabetes had clinical improvement after immunosuppressive treatment and only one patient had disease progression after the treatment. Rapidly progressive motor weakness and clinically improvement after immunosuppressive treatment were support the diagnosis of CIDP in diabetes patients. CIDP with diabetes patients were common in the present study and were not different from previous studies, which showed 9 to 26%[22]. However, identifying CIDP in diabetes patients is difficult because neuropathy in diabetes patients are varies and DPN can demonstrated demyelinating features on nerve conduction studies as well as raised CSF protein. To date, there is no established criterion for the diagnosis of CIDP in diabetes patients[23]. The present study could demonstrate CIDP in diabetes patients by using the clinical of classic CIDP with rapidly progressive motor weakness and diagnostic criteria in the EFNS/PNS guideline. This combination may imply to identifying CIDP in diabetic patients.

The clinical courses in the present study were varied. The majority of patients (71.4%) had disease duration prior to first evaluation more than eight weeks, but acute (less than 4 weeks) or subacute (4 to 8 weeks) disease onset can be found. These patients had diagnosis of AIDP and SIDP before, but their clinical

courses were relapsing or progressive with duration more than 8 weeks. Acute onset CIDP is common, and up to 20% of CIDP patients presented with acute onset in previously published studies[24,25]. Distinguishing acute onset CIDP from AIDP is necessary because treatment and prognosis are difference. Long-term immunosuppressive treatment is needed in acute onset CIDP patients. To date, there is no established criterion for distinguishing acute onset CIDP in the early phase of disease from AIDP. In the present study, autonomic symptoms, respiratory muscle involvement, and facial or cranial nerve involvement were rarely presented. These might be clues to distinguish acute onset CIDP from AIDP[26,27].

The efficacy of immunosuppressive treatment in CIDP including corticosteroid, IVIg, and plasmapheresis have been reported in many studies, but long-term prognosis of CIDP patients after treatments is still unclear[11-13]. In the present study, patients received various regimens of immunosuppressive treatment. The prognosis was generally favorable with 71.4% having mild disability after treatment. Nevertheless, there were 7.9% of patients who had severe disability, despite receiving immunosuppressive treatment. Out of the 63 patients, 68.3% got disease remission or were stable and 31.7% had disease relapse. Patients in the disease relapsing group were more likely to present with acute onset CIDP mimicking AIDP and had more severe disability. The mode of disease onset may be a predicting factor for the responsiveness to treatment. These were reported in a previous study, but further systematic study is still needed[28].

In summary, clinical characteristics, phenotypes, electrophysiological findings, and treatment response of CIPD in Thai patients were not very different from previously published studies in western and oriental populations. The mode of disease onset may help to predict response to immunosuppressive treatment in CIDP patients.

What is already known on this topic?

The prevalence and incidence of CIDP have greatly varied among publications. These could be related to genetic predisposition, using different clinical spectrum, and diagnostic criteria.

Typically, the disease onset of CIDP are chronic but acute or subacute (4 to 8 weeks) disease onset can be found. To date, there is no established criterion for distinguishing acute onset CIDP in the early phase of disease from AIDP but it is necessary because treatment and prognosis are difference. Absence of the autonomic symptoms, respiratory muscle weakness and facial or cranial nerve involvement might be clues to distinguish acute onset CIDP from AIDP.

CIDP is a treatable disease, usually response to immunosuppressive treatment but long-term prognosis of CIDP is still unclear.

What this study adds?

The demographic, characteristics of polyneuropathy, clinical phenotypes, electrodiagnostic features, and CSF profiles of CIDP in Thai patients were not different from the western and oriental populations.

The treatment response to immunosuppressive therapies are generally favorable. Nevertheless, some patients still had severe disability or disease relapse. Severe disease disability at onset and acute onset mimicking AIDP may be a predicting factor for the poor response to treatment.

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Potential conflicts of interest

None.

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ลักษณะทางคลินิกและการตอบสนองต่อการรักษาของผู้ป่วย chronic inflammatory demyelinating polyradiculoneuropathy

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ภูมิหลัง: *อุบัติการณ์ ความชุกของโรค อาการทางคลินิก และการตอบสนองต่อการรักษาของโรค chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) มีความหลากหลาย และแตกต่างกันในแต่ละประเทศ ซึ่งปัจจุบันยังไม่มีข้อมูลการศึกษาทางคลินิกของโรคกลุ่มนี้ในประเทศไทย*

วัตถุประสงค์: *1) เพื่อศึกษาลักษณะอาการทางคลินิก ผลการตรวจเส้นประสาทด้วยไฟฟ้า และการตอบสนองต่อการรักษาในผู้ป่วย CIDP และ 2) เพื่อหาปัจจัยชี้วัดการพยากรณ์โรคในผู้ป่วย CIDP*

วัสดุและวิธีการ: *ทำการรวบรวมและศึกษาข้อมูลจากเวชระเบียนของผู้ป่วย CIDP ที่ได้รับการวินิจฉัย ตรวจรักษา และติดตามอาการที่สถาบันประสาทวิทยาระหว่าง วันที่ 1 มกราคม พ.ศ. 2551 ถึง 31 ธันวาคม พ.ศ. 2557*

ผลการศึกษา: *จากการศึกษาผู้ป่วย CIDP จำนวน 63 ราย พบว่าผู้ป่วยผู้ชายพบบ่อยกว่าผู้หญิง (ชาย:หญิง 1.3:1) อายุเฉลี่ย 47.7 ปี มีอาการเฉลี่ย 5 เดือน ก่อนมาพบแพทย์ ระยะเวลาติดตามอาการหลังได้รับการรักษาเฉลี่ย 26.8 เดือน ผู้ป่วยร้อยละ 19 มีโรคเบาหวานร่วมด้วย ผู้ป่วยร้อยละ 76.2 มีลักษณะอาการของโรคเข้าได้กับ classic CIDP ร้อยละ 19 เข้าได้กับ DADS และร้อยละ 4.8 เข้าได้กับ MADSAM สำหรับระยะเวลาของโรคก่อนพบแพทย์ ผู้ป่วยร้อยละ 15.9 มาพบแพทย์ในระยะเวลาน้อยกว่า 4 สัปดาห์ (acute onset CIDP mimicking AIDP) ร้อยละ 12.7 มาพบแพทย์ในระยะ เวลาระหว่าง 4 ถึง 8 สัปดาห์ (SIDP) ผู้ป่วยส่วนใหญ่มีอาการ symmetrical, sensorimotor polyneuropathy ร่วมกับภาวะ hyporeflexia สำหรับอาการของระบบประสาทอัตโนมัติ ภาวะหายใจล้มเหลว ความผิดปกติของการพูดหรือกลืน ความผิดปกติของการกลอกตา หนังตาตก และกล้ามเนื้อลีบ พบได้น้อยในการศึกษานี้ ผู้ป่วยส่วนใหญ่ตอบสนองดีต่อ immunosuppressive treatment ในกลุ่มผู้ป่วยที่มีอาการกำเริบซ้ำหลังได้รับการรักษา พบว่าระยะเวลาก่อนมาพบแพทย์สั้นกว่าผู้ป่วยที่ไม่มีอาการกำเริบซ้ำ (2 vs. 6 เดือน) และร้อยละ 40 ของผู้ป่วยที่มีอาการกำเริบซ้ำ เป็นผู้ป่วยมาพบแพทย์เพื่อทำการตรวจวินิจฉัยครั้งแรกภายใน 4 สัปดาห์ หลังจากเริ่มมีอาการ*

สรุป: *อาการทางคลินิก อาการแสดงของโรค ผลการตรวจเส้นประสาทด้วยไฟฟ้า และการตอบสนองต่อการรักษาของโรค CIDP ในผู้ป่วยไทย ไม่มีความแตกต่างจากชาวเอเชีย และชาติตะวันตก ระยะเวลาของโรคก่อนพบแพทย์อาจช่วยพยากรณ์ผลการตอบสนองต่อการรักษาด้วย immunosuppression*