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Long-term follow-up of relapse and remission of CIDP in a Chinese cohort

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ABSTRACT

Objective We aim to describe the long-term outcome of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) after immune treatment in a Chinese cohort.

Methods Between March 2015 and March 2023, 89 patients fulfilling the criteria for CIDP were followed up for a median of 22 months after treatment. Nine had positive antibodies against nodal-paranodal cell-adhesion molecules. Patients were treated according to clinical requirements with prednisone, intravenous immunoglobulin (IVIg) and/or immunosuppressant.

Results A total of 78/89 patients had decreased inflammatory neuropathy cause and treatment (INCAT) scores at the last follow-up. For CIDP patients treated with steroids, 35 were stable without relapse after cessation or with a small maintenance dose; 2 relapsed at a high dose (20 mg/day); 15 relapsed at a low dosage (<20 mg/day) and 11 did not respond. The INCAT before treatment was significantly lower in those without relapse (median INCAT 2 vs 3, p=0.030). IVIg was effective in 37/52 CIDP patients. 28 CIDP patients and 4 autoimmune nodopathy patients were treated with immunosuppressants. The average INCAT was 3.3±1.9 before and 1.9±1.3 after immunosuppressant treatment (p=0.001) in CIDP.

Conclusion The long-term prognosis of CIDP patients was generally favourable. Nearly half of our patients treated with steroid were stable without relapse after cessation or with a small maintenance dose. The risk of relapse was higher in those with high INCAT. We recommend slowly tapering prednisone based on clinical judgement.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ The long-term clinical outcome of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) after treatment was only described in a few articles. We intend to describe the long-term clinical outcome following maintenance treatment in a large cohort of CIDP patients in China.

WHAT THIS STUDY ADDS

⇒ Our results showed that the long-term prognosis of CIDP patients was generally favourable. Nearly half of our patients treated with steroid were stable without relapse after cessation or with a small maintenance dose. The risk of relapse was higher in those with higher INCAT.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Our study provides the clinical outcome of CIDP following immune treatment. The traditional treatment showed favourable effects.

INTRODUCTION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is the most common chronic immune-mediated inflammatory polyneuropathy. Austin described a group of patients with recurrent polyneuropathies with a dramatic response to steroids and relapse induced by steroid withdrawal.[1] Dyck *et al* described the first randomised controlled trial (RCT) in CIDP, showing that prednisone caused significant improvement over no treatment. RCTs showed that intravenous immunoglobulin (IVIg), subcutaneous immunoglobulin (SCIg) and plasma exchange (PE) were efficacious as treatments for CIDP.[2–10] Case series have provided evidence for the efficacy of immunosuppressants, including cyclophosphamide (CTX), azathioprine (AZA), mycophenolate mofetil (MMF), ciclosporin A (CsA) and rituximab (RTX). Based on these studies and clinical evidence, guidelines strongly recommend IVIg, corticosteroid or PE as initial treatment and IVIg, subcutaneous immunoglobulin or corticosteroids for maintenance treatment.[11] However, the long-term clinical outcome of CIDP after treatment was only described in a few articles, including Bus *et al* from the Netherlands[12] and Kuwabara *et al* from Japan.[13] We intend to describe the long-term clinical outcome following maintenance treatment in a large cohort of CIDP patients in China.

MATERIALS AND METHODS

Patients

Between March 2015 and March 2023, consecutive patients with CIDP were recruited prospectively. For the diagnosis of definite CIDP, including typical CIDP and CIDP variants, we used the diagnostic criteria proposed by the Joint Task Force of the European Federation of Neurological Societies and the Peripheral Nerve Society.[14]

Clinical assessment

All patients underwent neurological examination and neurophysiological studies. Functional disability was assessed with the inflammatory neuropathy cause and treatment (INCAT) disability score[15] and Hughes functional grading scale.[16] Patients were treated according to clinical requirements. Patients with greater disability were advised to use IVIg as induction treatment. IVIg was given at 0.4 g/kg/day for 5 days. IVIg treatment is defined as effective if INCAT improves 1 or more in the fourth week after treatment. Prednisone was given at 60 mg/day slowly tapered to 40 mg at 3 months, slowly tapered to 20 mg/day or less over 3 months, and then maintained at a low dose for 1–3 years. Prednisone treatment is effective if INCAT improves 1 or more at 90 days after treatment. If steroids were ineffective or effective but unable to reduce below 20 mg/day and IVIg was unavailable or ineffective, an immunosuppressant was given. AZA was given 2–3 mg/kg/day; CTX 1–3 mg/kg/day; MMF 1–2 g/day; CsA 3–6 mg/kg/day; tacromus 1–2 mg/day; rituximab 100 mg on the first day and 500 mg on the second day.

Electrophysiological studies

Electromyography (EMG) and nerve conduction studies (NCSs) were performed with a Nicolet EMG machine (CareFusion, Middleton, Wisconsin). The room temperature was maintained to ensure that the patients' skin temperature remained above 31°C. Motor NCSs were performed in all participants on the median, ulnar, fibular and tibial nerves with percutaneous supramaximal nerve stimulation while recording the compound motor action potentials (CMAP) with 10 mm disk electrodes. F waves of the median and tibial nerves were performed in all participants with repetitive supramaximal stimulations at a rate of 1 Hz. Bilateral nerves were studied in all cases. The median nerve was stimulated at the wrist, elbow and axilla with recording from the abductor pollicis brevis; the ulnar nerve was stimulated at the wrist, below the elbow, above the elbow and at the axilla with recording from the abductor digiti minimi. Measurements included distal motor latency, motor conduction velocity, CMAP amplitude (baseline to negative peak), and area and duration of the negative wave.

Orthodromic sensory NCSs were performed on the median, ulnar, medial plantar and superficial fibular nerves. Conventional needle EMG was performed in the tibialis anterior and extensor digitorum communis muscles.

Antibody testing

Nodal and paranodal (including anti-NF155, anti-CNTN1, anti-Caspr1, anti-NF140/186) antibodies were tested using a cell-based assay.

Statistical analysis

Statistical analysis was performed in SPSS V.10.01 (SPSS). Patient characteristics were explored using descriptive statistics. The Kolmogorov-Smirnov test was used to test the normality of age, disease duration, INCAT score and steroid dosage, which showed a non-normal distribution.

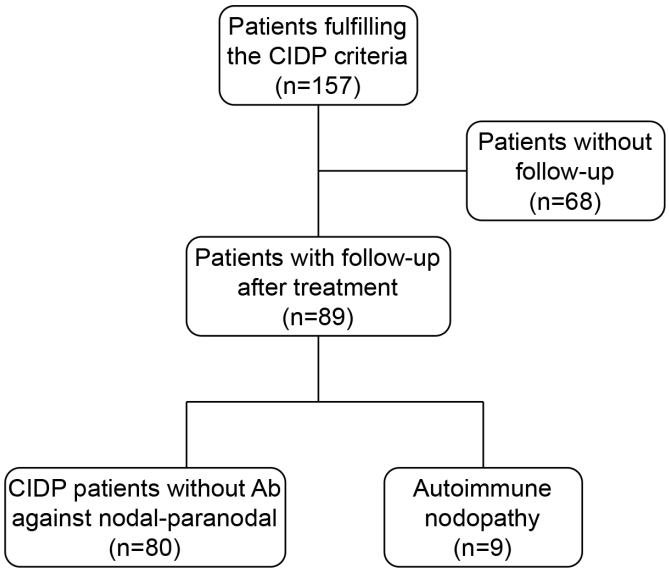

Figure 1 Flow chart of inclusion. CIDP, chronic inflammatory demyelinating polyradiculoneuropathy.

The Wilcoxon test was used for comparisons between non-normally distributed variants. The χ^2 test was used for comparisons between categorical variables. Two-sided p values were calculated for all analyses; a $p \leq 0.05$ was considered significant.

RESULTS

Demographic features of patients

A total of 157 patients were recruited, of whom 89 were followed up after treatment (figure 1). The median age was 45 years old (IQR, 29–59.5, ranging 12–80). There were 55 males and 34 females. The median disease duration at the beginning of treatment was 6 months (IQR 4–24) and that at the last follow-up was 33 months (IQR 20–62). Nine patients had positive antibodies against nodal-paranodal cell-adhesion molecules (3 NF155+, 1 NF186+, 1 NF155+NF186+, 3 CNTN1+ and 1 Caspr1+). Among the 80 other CIDP patients, clinical subtypes included 53 typical CIDP, 6 multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), 11 distal acquired demyelinating symmetric neuropathy (DADS), 8 pure motor CIDP and 2 pure sensory CIDP.

Of all 89 patients, after treatment with steroids, IVIg and/or immunosuppressants, 84 patients showed clinical improvement while 5 patients showed deterioration or no response. The median INCAT before treatment was 3 (IQR 2–3.25) and that at the last follow-up was 1 (IQR 0–2). At the last follow-up, only 4 patients had increased INCAT, 78 had decreased INCAT and 7 had unchanged INCAT.

Treatment with steroids

Of the 80 CIDP patients without antibodies against nodal-paranodal cell-adhesion molecules, 79 were treated with steroids. Steroids were effective in 69 patients and

Table 1 General information about the four groups of patients with different responses to steroids

	n	Disease duration when treatment was started/month	Duration of steroid treatment/month	Follow-up duration after beginning of steroid treatment/month	INCAT before treatment	CSF-Protein g/L	Highest hughes
Group 1a	9	5 (3–9.5)	25 (19.5–31)	30 (25–53)	2 (1–2.5)	2.3 (0.9–3.7)	1.5 (1–2)
Group 1b	26	9 (4–24)	18.5 (11–34.5)	18.5 (11–34.5)	2 (1.5–4)	2 (2–3)	1.0 (0.6–1.8)
Group 2	15	7 (4–17)	21 (12–45)	39 (20–69)	3 (2–4)	0.8 (0.5–1.4)	2 (2–3)
Group 3	2	29.5	14.5	14.5	2.5	1.1	2
Group 4	11	24 (4–60)	12.5 (3.75–26)	14 (8.8–35)	3 (2–3)	0.7 (0.5–2.3)	2 (2–3)

Group 1a: patients with no relapse after cessation of steroid; Group 1b: patients with no relapse with small maintaining steroid; Group 2:, patients who had relapse with small steroid maintaining dosage (<20 mg/day); Group 3: patients who had relapse when steroid dosage was >20 mg/day; Group 4: Steroid was ineffective. Figures were median (IQR).
CSF, cerebral spinal fluid; INCAT, The Inflammatory Neuropathy Cause and Treatment Disability Score.

ineffective in 10. All nine patients with autoimmune nodopathies were treated with steroids.

The responses to steroids were classified into four groups: group 1: No relapse after cessation of steroids or a small dosage of steroids (<20 mg/day) without other treatment. Patients treated with immunosuppressants were not included in this group. Group 2: Relapse with small steroid maintenance dosage (<20 mg/day). Group 3: Relapse when the steroid dosage was more than 20 mg/day. Group 4: Steroids were ineffective. The general information about the four groups is summarised in table 1.

In group 1, nine patients were stable without relapse after cessation of steroids. They were treated for a median time of 25 months (IQR 19.5–31) and were followed up for a median time of 4 months (IQR 1.5–20) after the cessation of steroids. 26 patients were stable with a small maintenance dose of steroids (<20 mg/day) without other treatment. The median maintenance dosage was 7.5 mg (IQR 5–15 mg; range 2.5–20 mg).

In group 2, 15 patients relapsed at a low steroid dosage (<20 mg). Nine relapsed once, and six relapsed twice or more. Among them, immunosuppressants were added to seven patients. Others had steroid dosage increased. The average steroid dosage at which relapse occurred was 6.2±5.4 mg (ranging from 0 to 15 mg).

In group 3, two patients relapsed at a high steroid dosage (20 mg/day). AZA was added to both patients.

In group 4, steroids were ineffective in 11 patients. Immunosuppressants were added in seven of them. IVIg was used in the other four patients.

We compared group 1 and group 2. Disease duration when steroid treatment started was not significantly different (median duration 6.5 vs 7 months, p=0.991 for M-W test). The INCAT before treatment was significantly lower in those without relapse (median INCAT 2 vs 3, p=0.030 for M-W test). The cerebrospinal protein and highest Hughes score were not significantly different (p=0.071 and 0.341, respectively, with the M-W test).

The clinical subtypes and steroid treatment response are illustrated in table 2. The percentage of relapse was higher in MADSAM than in typical CIDP (p=0.161 for Pearson's χ^2 test) and lower in DADS or pure motor CIDP than in typical CIDP (p=0.454 and p=0.385, respectively, for Pearson's χ^2 test); however, both were not statistically significant.

For the nine patients with positive antibodies against nodal-paranodal cell-adhesion molecules, all were responsive to steroids. Two patients were stable with small maintenance doses of steroids (7.5 and 12.5 mg/day, respectively) without other treatment. Two patients relapsed at a low steroid dosage (5 mg and 12.5 mg).

Severe adverse effects of steroids occurred in two patients. One had femoral head necrosis, so steroids were stopped, and AZA was added. The other had facial

Table 2 The response to steroids in different clinical subtypes

	Typical	MADSAM	Distal CIDP	Pure sensory CIDP	Pure motor CIDP	Total
No relapse after cessation of steroid or small maintaining dosage	26	1	6	1	2	36
Relapse with small dosage of steroid (<20 mg)	10	2	1	1	0	14
Steroid was ineffective	3	3	2	0	3	11
	39	6	9	2	5	61

CIDP, chronic inflammatory demyelinating polyradiculoneuropathy; MADSAM, multifocal acquired demyelinating sensory and motor neuropathy.

infection, so steroids were tapered quickly to 10 mg/day, and CTX was added.

Treatment with immunosuppressants

28 CIDP patients without nodal-paranodal antibodies were treated with immunosuppressants. AZA was given to 16 patients. CTX was given to nine patients. MMF was given to 10 patients. CsA was given to two patients. Tacrolimus was given to two patients. Six patients were treated with one immunosuppressant and later switched to another because of side effects or irresponsiveness. There were 16 males and 12 females. The median age was 51.5 years old (IQR 33–60). The average disease duration when immunosuppressants were added was 33.9±32.4 months. Clinical subtypes included 17 typical CIDP, 4 MADSAM, 1 DADS, 1 pure sensory and 5 pure motor. The average immunosuppressant treatment duration was 17.3±11 months. 15 were responsive, 5 were unresponsive and 1 was responsive at first and unresponsive later. The average INCAT was 3.3±1.9 before immunosuppressant treatment and 1.9±1.3 after immunosuppressant treatment (p=0.001, Wilcoxon test). The annual recurrence rate was 0.8±1.8 before immunosuppressant treatment and 0.1±0.3 after immunosuppressant treatment (p=0.016, Wilcoxon test).

Among the nine patients with antibodies against nodal-paranodal cell-adhesion molecules, four used immunosuppressants. CTX was given in one patient, AZA in one, RTX in one and AZA and RTX in one. Immunosuppressants were effective in all four patients. The average INCAT was 3.3±0.6 before immunosuppressant treatment and 1±1 after immunosuppressant treatment. The annual recurrence rate was 0.3±0.6 before immunosuppressant treatment and 0 after immunosuppressant treatment.

Azathioprine

Two patients stopped AZA due to side effects, one because of reduced white blood cells and the other because of elevation of liver enzymes.

Among the 14 patients who used AZA, 8 were responsive, 2 were unresponsive, 1 was responsive in the first year but unresponsive later and 3 did not have enough follow-up time. The average INCAT was 2.5±1.6 before AZA and 1.5±1.4 after AZA (p=0.016, Wilcoxon test, figure 2). The annual recurrence rate was 1.3±2.3 before AZA and 0.2±0.3 after AZA (p=0.042, Wilcoxon test).

Other immunosuppressants

Among the five patients who used CTX, four responded to the treatment and one did not respond (figure 2).

Among the five patients who used MMF, all responded to the treatment (figure 2).

Treatment with IVIg

IVIg was effective in 37/52 CIDP patients. Since it is expensive and not covered by general medical insurance, most patients responsive to IVIg were treated with steroids as maintenance treatment. Only one patient recovered after treatment with one round of IVIg and did not relapse at the last follow-up.

For patients with autoimmune nodopathies, IVIg was administered in four patients, and they did not respond.

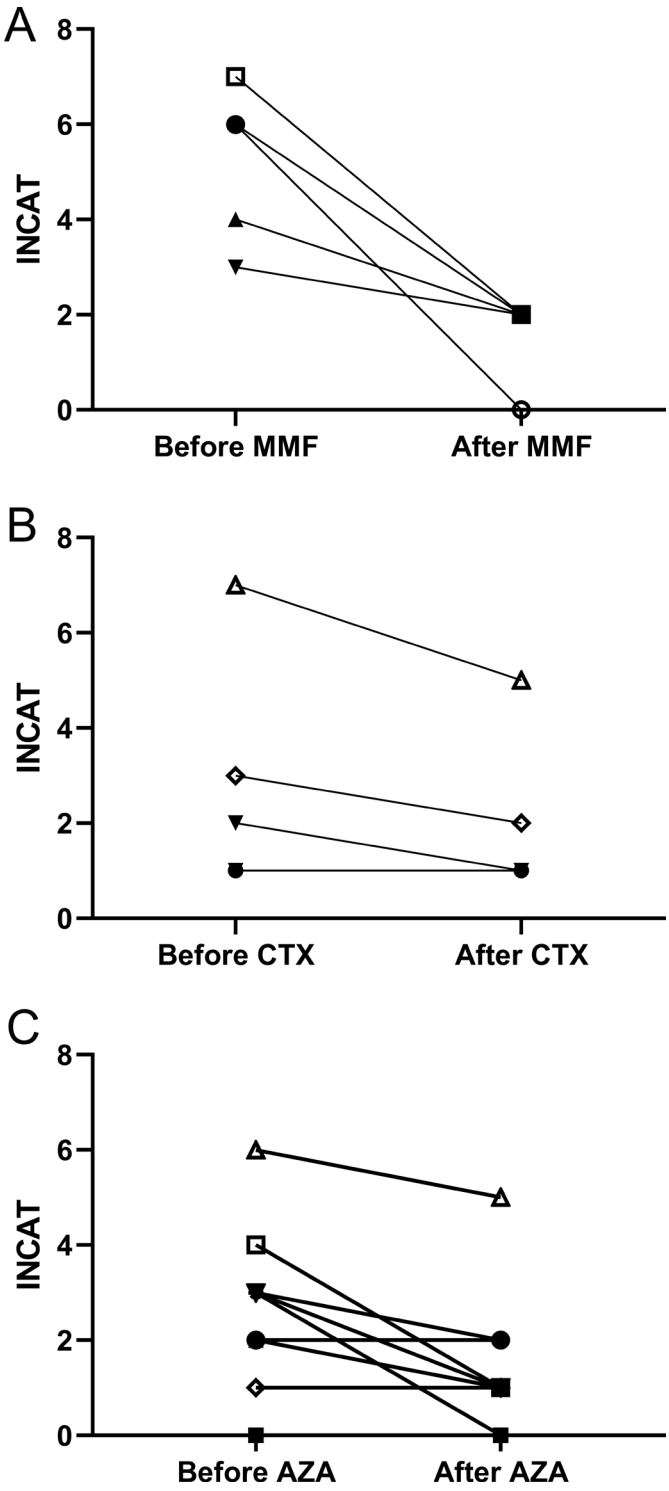

Figure 2 The inflammatory neuropathy cause and treatment (INCAT) disability score in (A) 11 patients before and after azathioprine (AZA) treatment. (B) Five patients before and after cyclophosphamide (CTX). (C) Five patients before and after AZA. MMF, mycophenolate mofetil.

DISCUSSION

Immune treatments are proven to be useful in CIDP, including steroids, IVIg, PE and immunosuppressants. However, the long-term clinical outcomes following treatment, including relapse rate, remission rate and clinical features that might indicate clinical prognosis, were only described in small cohorts in a few articles. The strengths of this study on real-world data are the focus on the large patient number and the relatively long follow-up. The long-term prognosis of CIDP patients following immune treatment, especially steroids, was described.

According to the study of Kuwabara *et al*, the long-term prognosis of CIDP patients was generally favourable. 26% of patients had complete remission, and 61% had partial remission with (26%) or without (34%) immune treatment.[13] Our follow-up study showed a favourable treatment response in Chinese CIDP patients. Most patients were responsive to immune treatment, and only 6% of patients showed deterioration or no response despite treatment. The long-term prognosis was also generally favourable. At the last follow-up, the INCAT decreased in 87% of patients. 11% of patients were stable without relapse after cessation of steroids, and 33% of patients were stable with only a small maintenance dose of steroids. 35% of patients required immunosuppressants.

Steroids were used in most patients as maintenance treatment. Patients showed different responses to long-term steroid treatment, including no relapse after steroid cessation or with a small maintenance dosage of steroids, relapse at a high steroid dosage, relapse at a small steroid dosage and steroid ineffectiveness. The INCAT before treatment was significantly lower in those without relapse, indicating a higher risk of relapse in those with more clinical impairment before treatment. Regarding clinical subtypes, the relapse rate was higher in MADSAM and lower in pure motor and distal CIDP than in typical CIDP. However, this was not significantly different and should be confirmed with larger studies. These results might indicate that for patients with higher neurological function impairment and the MADSAM subtype, the risk of relapse is higher. Steroids might be reduced more slowly, and immunosuppressants are more likely to be added. In distal CIDP and pure motor CIDP responding to steroids, however, the risk of relapse is lower. Two out of the five pure motor CIDP patients responded to IVIg, indicating that not all pure motor CIDP must be treated with IVIg. This was consistent with previous studies.[17–20] For the nine patients without relapse after cessation of steroids, the disease duration when treatment started was short, and INCAT before treatment was lower compared with those who had relapse. This indicates that earlier treatment and less severe clinical impairment might be related to a better prognosis.

Information about the best exact tapering strategy of steroids for CIDP is incomplete. Current recommendations are based on published case reports and personal experience. Wertman *et al* found that lowering steroids must be very gradual and that treatment for less than 6 months and rapid tapering off from steroids may increase the risk of relapse.[21] van Schaik *et al* recommended starting prednisolone 60 mg/day for 4–8 weeks and tapering the dose over 52–104 weeks.[22] Odaka recommended slowly reducing the steroid dose over a 12-month period.[23] In our study, those who were stable without relapse had been treated for approximately 2 years before the cessation of steroids. Based on our results and experience, we recommend starting prednisone 60 mg/day or 1 mg/kg/day and slowly tapering to 20 mg/day in 6 months, then slowly tapering and maintaining for approximately 2 years. If relapse occurred when steroids were more than 20 mg/day, immunosuppressants could be added. IVIg could also be used during severe relapse.

The efficacy of immunosuppressants in treating CIDP was studied in randomised trials in methotrexate, interferon beta 1a, fingolimod and AZA.[24–27] The results, however, were unfavourable. Case series have provided evidence for the usage of AZA, MMF, CTX, CsA and rituximab in patients insufficiently responding or refractory to conventional treatment.[11] Our results provide further evidence for the usage of these immunosuppressants. Immunosuppressants, including AZA, CTX, MMF, CsA and tacrolimus, were given to 28 patients. Most patients (71%) were responsive. The average INCAT and average year relapse rate were dramatically reduced after adding immunosuppressants. AZA was most often used due to its relatively high response rate, low frequency of side effects (2/16) and low price.

Autoimmune nodopathies often have specific clinical characteristics. Case reports showed no or poor response to IVIg treatment, partial response to steroids and possible response to rituximab.[28–32] In our study, 9 out of 89 patients had positive antibodies against nodal-paranodal cell-adhesion molecules. All patients were responsive to steroids. For the five patients who had been followed up for a median of 16 months (IQR 8.5–20.5 months), two were stable with a small maintenance dose of steroids, two relapsed at a low steroid dosage and CTX was added to the other. None of the four patients who used IVIg were responsive. 44% (4/9) of patients with nodal-paranodal antibodies were treated with immunosuppressants, compared with 35% of CIDP patients. CTX, AZA and RTX were used, and all were effective. Our results further proved the poor response to IVIg and good response to steroids and immunosuppressants in autoimmune nodopathies.

Our study has some limitations. First, it is a real-world study of CIDP treatments. Not all patients were treatment-naive, disease duration varied among patients and the treatment option of immunosuppressant was based on the patients' conditions, including economic status, fertility requirements, etc. Second, a large proportion of patients were lost to follow-up. 17 patients were newly included and did not have enough time to follow-up. Our hospital is a tertiary hospital in the capital. A great portion of our patients came from other cities all around the country. Distance and inconvenience might be the main reason for the patients to lose to follow-up. However, their loss to follow-up might have effect on the study's validity. Third, some patients were followed up for less than 6 months.

In conclusion, our results showed that the long-term prognosis of CIDP patients was generally favourable. Clinical disability recovered fully or partially in most patients. 44% of

patients were stable without relapse after cessation or with a small maintenance dose of steroids. Some patients relapsed and required immunosuppressants. The risk of relapse was higher in those with higher INCAT. According to our data, prednisone slowly tapered based on clinical judgement was recommended. If frequent relapse occurs despite slow tapering, immunosuppressants, including AZA, CTX or MMF, should be considered.

Contributors JN: clinical studies, data acquisition and analysis, statistical analysis, manuscript preparation; LZ: data acquisition; NH: statistical analysis; LC: concept; ML: concept, design, manuscript editing and review. JN is responsible for the overall content as guarantor.

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Competing interests None declared.

Patient consent for publication Consent obtained directly from patient(s).

Ethics approval This study involves human participants and the ethics committee of Peking Union Medical College Hospital approved our study protocol. The ethics approval number was I-23PJ381. Participants gave informed consent to participate in the study before taking part.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data are available on reasonable request.

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