Real-world treatment patterns in adults with chronic inflammatory demyelinating polyneuropathy in the United States

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Introduction and Purpose

- Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare, immune-mediated disorder of the peripheral nervous system characterized by sensory impairment, progressive muscular dysfunction, and fatigue.[1] In the United States (US), the annual prevalence rate of CIDP is 8.9 per 100,000 persons.[2]
- The primary treatment modalities for CIDP include immunoglobulin (Ig), corticosteroids, and plasma exchange (PLEX).[1] Factors such as heterogeneity in disease severity, presence of comorbidities, potential adverse effects, and access can play a key role in the selection of treatment.[1,3,4]
- While real-world treatment patterns in CIDP have been explored,[5-7] variability in methodologies, such as population selection or databases used, may confound the representation of burden in CIDP. Thus, there is a need to examine CIDP treatment patterns among a robust sample to elucidate care delivery gaps, identify vulnerable patient populations, and develop effective strategies to further improve health outcomes.
- The aim of this study was to evaluate treatment patterns in patients with CIDP in the US.

Methods

- A retrospective cohort study was conducted using Komodo Health (a US-based claims database containing complete medical and prescription claims information from 150 payers across all geographic regions in the US) from January 2016 to December 2020 (**Figure 1**).

Figure 1. Study design

Key patient selection criteria

- ≥2 claims with CIDP diagnosis, ≥30–≤365 days apart (first observed CIDP diagnosis was considered as index date)
- Continuous enrollment ±1 year pre- and post-index date
- ≥1 nerve conduction test[a] present either after the index date and before another CIDP diagnosis, or ≤90 days before the index date
- Age ≥18 years on index date
- Closed claims

Study variables

- Patient baseline characteristics (pre- or at index date)

Exclusion criteria

- ≥2 of the same exclusionary diagnoses during 2-year study period[b]

Study outcomes

- CIDP-related treatments: claims, dose, episodes, frequency, etc, in overall population and by subgroups in the 1-year post-index period

Statistical analysis

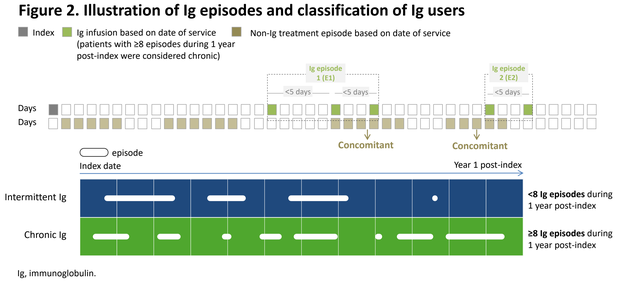
- Descriptive statistics were used to analyze patient baseline characteristics and CIDP-related treatments

[a]A nerve conduction test requirement was added to increase the robustness and certainty of identifying patients with CIDP.
[b]Exclusionary diagnosis includes amyloidosis, amyotrophic lateral sclerosis, autoimmune hemolytic anemia, B12 deficiency, celiac disease, chronic lymphocytic leukemia, dermatomyositis, fibromyalgia, Guillain-Barre syndrome, familial neuropathy, human immunodeficiency virus, immune thrombocytopenic purpura, inclusion body myositis, bone marrow transplant, Kawasaki disease, multifocal motor neuropathy, multiple myeloma, multiple sclerosis, myasthenia gravis, necrotizing fasciitis, nonfamilial hypogammaglobulinemia, primary secondary immunodeficiency, sarcoidosis, organ transplant, systemic lupus erythematosus, toxic neuropathy, and cancer chemotherapy.
CIDP, chronic inflammatory demyelinating polyneuropathy.

Definitions associated with Ig utilization:

- **Ig:** Any claims indicating intravenous (IV) or subcutaneous (SC) Ig use were included.
- **Ig episode:** Among patients receiving Ig during 1 year post-index, one Ig episode was defined as a cluster of Ig infusions <5 days apart from one another (**Figure 2**). Patients were stratified into 2 cohorts based on the number of Ig episodes during 1 year post-index:
  - **Intermittent Ig:** <8 Ig episodes during 1 year post-index.
  - **Chronic Ig:** ≥8 Ig episodes during 1 year post-index.
- **Concomitant treatments:** Non-Ig treatments were classified into episodes based on date service. Any episodes that occurred concurrently with Ig episodes were considered concomitant treatments.

Figure 2. Illustration of Ig episodes and classification of Ig users



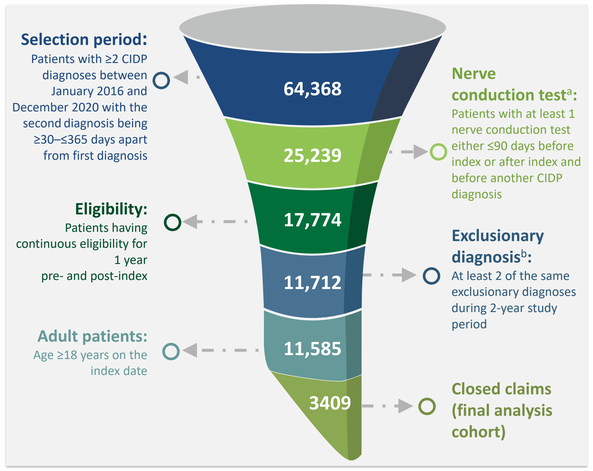
Ig, immunoglobulin.

Results

Patient selection

- Among the dataset, 3409 patients with CIDP were identified (**Figure 3**).

Figure 3. Patient selection



[a]A nerve conduction test requitement was added to increase the robustness and certainty of identifying patients with CIDP.
[b]Exclusionary diagnosis defined in study design.
CIDP, chronic inflammatory demyelinating polyneuropathy.

Baseline demographics and clinical characteristics

- Overall, baseline demographic and clinical characteristics were consistent with previous reports.[2] A significant proportion of patients with CIDP were predominantly between 41 and 65 years of age (**Table 1**).
- The most commonly observed comorbidities 1 year pre-index were diabetes without chronic complication (31% [n=1054]), chronic pulmonary disease (22% [n=753]), diabetes with chronic complication (21% [n=720]), cerebrovascular disease (15% [n=519]), and peripheral vascular disease (15% [n=514]; **Table 1**).

Table 1. Baseline patient demographics and clinical characteristics

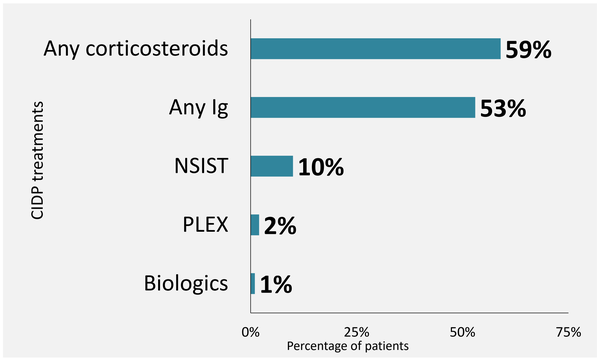
	N=3409
Age, years, mean (SD)	59.4 (13.9)
Distribution by age, n (%)	
18–40	335 (10)
41–65	1932 (57)
65+	1142 (33)
Gender, n (%)	
Male	2055 (60)
Female	1354 (40)
Race and ethnicity, n (%)	
Non-Hispanic Caucasian	1405 (41)
Hispanic	292 (8)
Non-Hispanic African American	162 (5)
Non-Hispanic Asian	25 (1)
Other/unknown	1525 (45)
CCI, mean (SD)	2.0 (2.2)
Insurance, n (%)	
Commercial	1680 (49)
Medicare	934 (27)
Medicaid	460 (13)
Other/multiple/unknown[a]	335 (10)
Treatments used 1 year prior to index, n (%)	
Steroids	1287 (38)
IVIg/IVIg or SCIg with combinations	521 (15)
IVIg or SCIg	136 (4)
Other combinations/treatments	120 (4)
IVIg	119 (4)
NSIST	33 (1)
PLEX	4 (0)
Biologics	2 (0)
Comorbidities, n (%)	
Diabetes without chronic complication	1054 (31)
CPD[b]	753 (22)
Diabetes with chronic complication	720 (21)
Cerebrovascular disease	519 (15)
Peripheral vascular disease	514 (15)
Renal disease	282 (8)
CHF	341 (10)
Any malignancy[c]	313 (9)

[a]Including self-insured, other/unknown, or dual-eligible.
[b]Including bronchitis, emphysema, asthma, chronic obstructive pulmonary disease, bronchiectasis, pneumoconiosis, and chronic drug-induced interstitial lung disorders
[c]Including lymphoma and leukemia, except malignant neoplasm of skin.
CCI, Charlson Comorbidity Index; CPD, chronic pulmonary disease; CHF, congestive heart failure; IVIg, intravenous immunoglobulin; NSIST, nonsteroidal immunosuppressive treatment; PLEX, plasma exchange; SCIg, subcutaneous immunoglobulin; SD, standard deviation.

Overall CIDP treatment utilization during 1 year post-index

- Among the 3409 patients, the majority were treated for CIDP during 1 year post-index (81% [n=2758]), while 19% (n=651) were untreated.
- The most common CIDP treatments used in the 1-year post-index period were corticosteroids (used at least once by 59% [n=2017]) followed by Ig (used at least once by 53% [n=1803]; **Figure 4**). Of the 2017 patients utilizing corticosteroids, 38% [n=771/2017] utilized steroids exclusively.

Figure 4. Proportion of patients who received CIDP treatments at least once during 1 year post-index (N=3409)



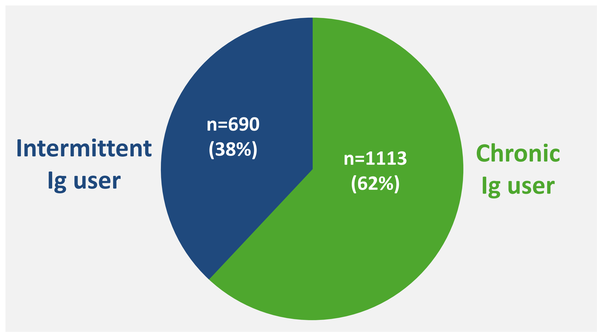
CIDP, chronic inflammatory demyelinating polyneuropathy; Ig, immunoglobulin; NSIST, nonsteroidal immunosuppressive treatment; PLEX, plasma exchange.

- Among the patients who received Ig treatment at least once during 1 year post-index, the majority (72% [n=1293/1803]) used Ig in combination with at least 1 other class of CIDP treatment.
- Of the patients who received Ig treatment in combination with other treatments, the most common was steroids, with 86% [n=1105/1293] utilizing steroids during 1 year post-index.

Ig utilization during 1 year post-index

- Of the 1803 patients who received Ig at least once in the 1-year post-index period, a significant proportion were chronic Ig users (62% [n=1113]; **Figure 5**).

Figure 5. Percentage of chronic Ig[a] users and intermittent Ig[b] users out of all patients receiving Ig in the 1-year post-index period (N=1803)

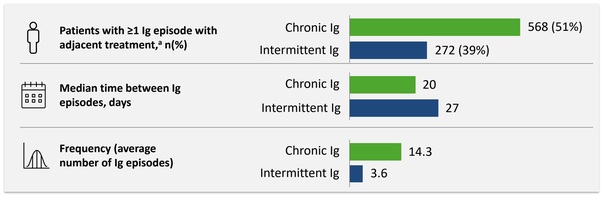


[a]Chronic IG users are patients with ≥8 Ig episodes during 1 year post-index.
[b]Intermittent IG users are patients with <8 Ig episodes in the 1-year post-index period.
Ig, immunoglobulin.

- Chronic Ig users were more likely to receive other concomitant CIDP therapies (51%) compared to intermittent Ig users (39%; **Figure 6A**).
- Oral steroids were the most commonly used concomitant treatment across Ig users, with its usage more prevalent among chronic Ig users versus intermittent Ig users (32% vs 20%; **Figure 6B**).

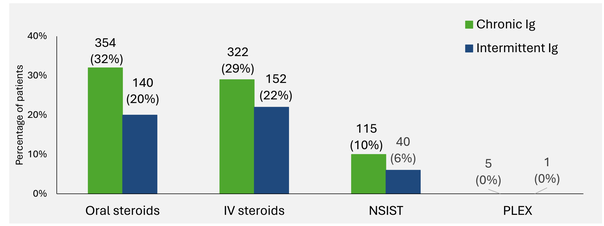
Figure 6. Concomitant treatment usage in chronic and intermittent Ig users

(A) Ig utilization patterns in chronic and intermittent Ig users



[a]Concomitant treatments are defined as non-IVIg treatments initiated and concomitant (based on date of service) from initiation of IVIg episode to the beginning of the next IVIg episode.
Ig, immunoglobulin; IVIg, intravenous immunoglobulin.

(B) Proportion of concomitant treatments in chronic and intermittent Ig users



Ig, immunoglobulin; IV, intravenous; NSIST, nonsteroidal immunosuppressive treatment; PLEX, plasma exchange.

- Of the 2017 patients who received steroids at least once in the 1-year post-index period, 60% (n=1204) received oral steroids and 49% (n=999) received IV steroids at least once.
- Among the 1204 patients who used oral steroids at least once, 1007 patients had dosing information available. Among the 1007 patients, time on oral steroid treatment was roughly 3 months on average, with 32% (n=320/1007) receiving >60 mg/day (**Table 3**).
- Even individuals using >60 mg/day oral steroids continued to use concomitant treatments, which was most commonly Ig (**Table 3**).

Table 3. Oral steroid usage in patients with dosage information available during 1 year post-index

	Overall N=1007 (100%)[a]	0–30 mg/day[b] N=352 (35%)	>30–60 mg/day N=335 (33%)	>60 mg/day N=320 (32%)
Annualized time on treatment, days				
Mean (SD)	89 (105)	113 (118)	75 (95)	81 (95)
Median (IQR)	35 (10–150)	60 (14–202)	30 (9–108)	30 (8–122)
Average daily dose mg/day				
Mean (SD)	60 (74)	17 (8)	45 (9)	122 (104)
Patients with usage of other treatments, n (%)				
Ig	581 (58)	185 (53)	210 (63)	186 (58)
NSIST	198 (20)	78 (22)	63 (19)	57 (18)
Biologics	26 (3)	9 (3)	8 (2)	9 (3)
PLEX	31 (3)	8 (2)	12 (4)	11 (3)

[a]Oral steroid patients were included if dosing information was available and could be calculated.
[b]Dosages are standardized based on prednisolone.
Ig, immunoglobulin; IQR, interquartile range; NSIST; nonsteroidal immunosuppressive treatment; PLEX, plasma exchange SD, standard deviation.

Conclusions

- **The mainstay treatments for CIDP were Ig and steroids during 1 year post-index.**
- **Among patients receiving Ig in the 1-year post-index period, the majority were chronic users, who had 8 or more Ig episodes.**
- **A substantial proportion of Ig users received concomitant CIDP treatments, with steroids being the most common.**
- **More than half of patients who received >60 mg/day oral steroids during 1 year post-index continued to use concomitant CIDP treatments, most commonly Ig.**
- **Further studies should identify any unmet needs (eg, clinical or economic) associated with the current CIDP management standard, in order to address any potential gaps in care.**

Limitations

- This study analyzed retrospective data from a claims dataset, which is limited in the capture of information such as disease severity or rationale for treatment selection.
- Given that the study population is drawn from the US, it may not fully represent the CIDP patient population in other geographical regions.

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