Poster P267

Subcutaneous Immunoglobulin (IgPro20) Dose Adjustments for Chronic Inflammatory Demyelinating Polyneuropathy Maintenance Therapy in Clinical Practice

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Introduction

- Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated neuropathy characterized by progressive or relapsing, symmetric, proximal and distal limb weakness, and impaired sensory function[1,2]
- The 2021 European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) CIDP guideline strongly recommends intravenous (IVIg) or subcutaneous (SCIg) immunoglobin for maintenance therapy of CIDP[2]
  - Compared with IVIg, SCIg offers more consistent immunoglobulin levels, which alongside reduced systemic adverse events, may improve quality of life[3]
- Using SCIg doses of 0.2 or 0.4 g/kg/week when transitioning from IVIg to SCIg has been shown to be effective for preventing disease relapse[4], and an equivalent mean weekly IVIg dose (1:1) is also recommended[2]
  - Optimal treatment uses the lowest effective dose tailored to a patient's response and needs
- There are limited data available on the clinical practicalities of individualizing SCIg

Objective

- To examine real-world evidence of the dosing of IgPro20 (immune globulin subcutaneous [human], 20% liquid, Hizentra®), in patients with CIDP, in clinical practice.

Methods

- This is a retrospective, non-interventional, anonymized study of 20 patients with CIDP from eight US centers who were treated with SCIg IgPro20 as maintenance therapy, following initial treatment with IVIg
- The study protocol and case study form were approved, and an Institutional Review Board waiver was obtained
- Data (demographics, diagnosis, treatment history, dosing information and decisions) were obtained from patient medical records
  - Inclusion criteria included being ≥18 years of age, a diagnosis of typical CIDP or a CIDP variant, positive response to IVIg induction treatment, and IgPro20 maintenance treatment for at least 6 months
  - Exclusion criteria included patients with concomitant autoimmune diseases, or any polyneuropathy of other causes
- The dose of IgPro20 at the initial transition from IVIg was noted, as well as any dose adjustments made for each patient

Results

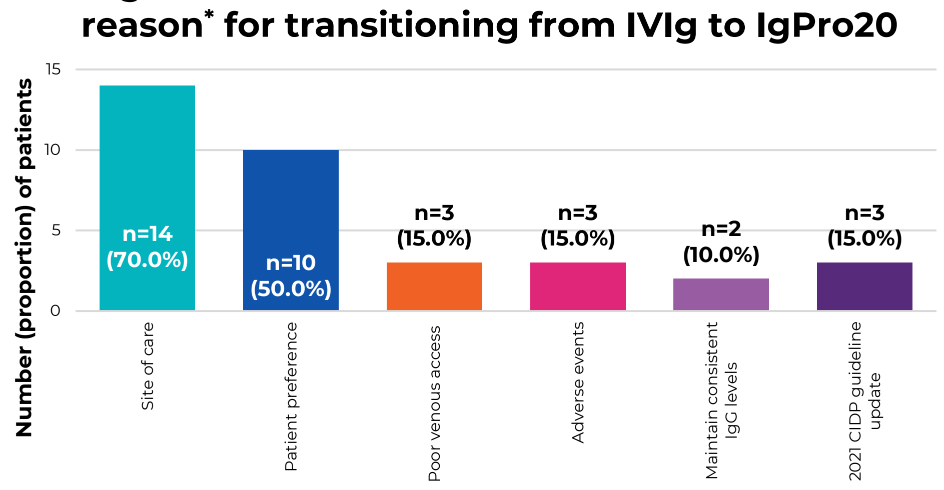
PATIENT DEMOGRAPHICS

- A total of 20 patients were included in the study cohort
- The mean (standard deviation [SD]) age of patients in the cohort was 59.5 (12.9) years and 55.0% (n=11) were male
- Fourteen patients (70.0%) were diagnosed with typical CIDP

PREVIOUS EXPERIENCE WITH IVIg

- Prior to transitioning to IgPro20, the mean (SD) length of time from CIDP diagnosis was 5.4 (6.2) years (n=19), the mean (SD) duration of IVIg use was 3.2 (4.4) years (n=20), and the mean (SD) equivalent weekly IVIg dose was 0.47 (0.42) g/kg/week (n=19)
- The most common reasons for transitioning to IgPro20 are described in **Figure 1**

Figure 1 – Site of care was the most common reason* for transitioning from IVIg to IgPro20

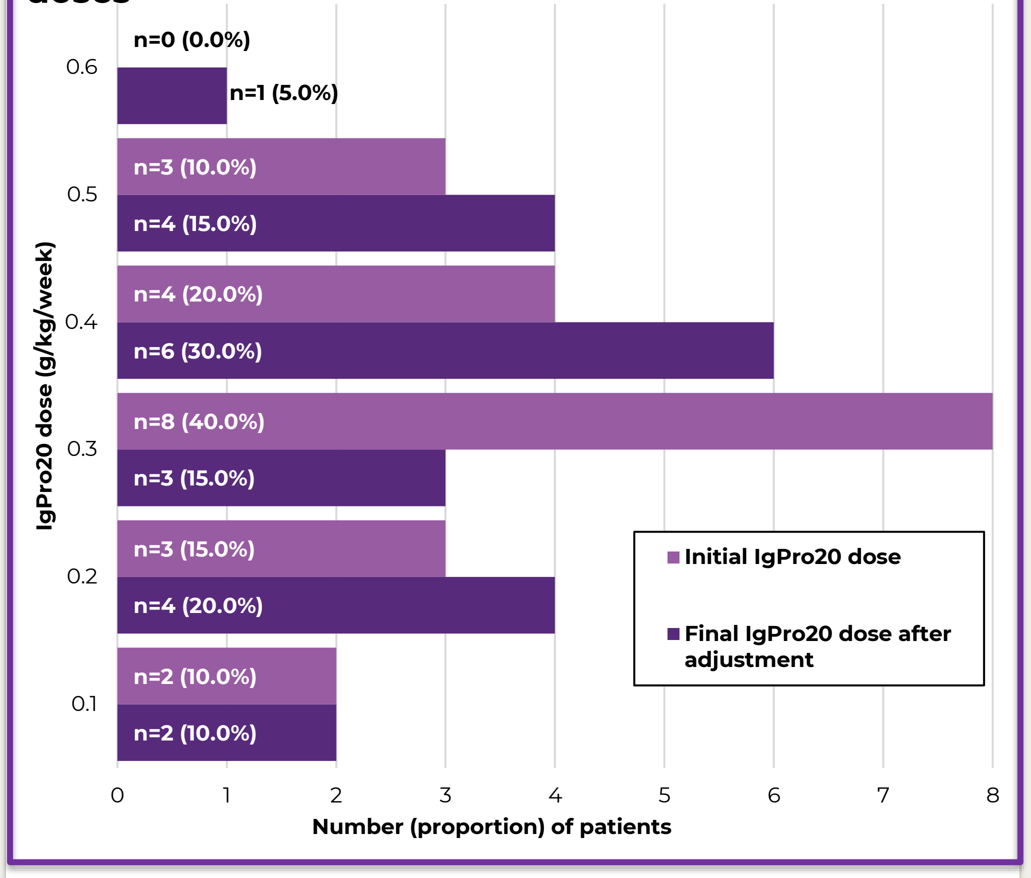


*Possible reasons for transition were not exclusive and multiple responses could be given. CIDP, chronic inflammatory demyelinating polyneuropathy; IgG, immunoglobulin G; IVIg, intravenous immunoglobulin.

INITIAL DOSE SELECTION OF IgPro20

- A large proportion of patients (n=8, 40.0%) transitioned from IVIg to SCIg on a dosing ratio of 1:1
- In this retrospective analysis, the mean (SD) initial dose of IgPro20 was 0.31 (0.10) g/kg/week, with 40.0% (n=8) of patients receiving an initial dose of 0.3 g/kg/week (**Figure 2**)

Figure 2 – Distribution of initial and adjusted IgPro20 doses

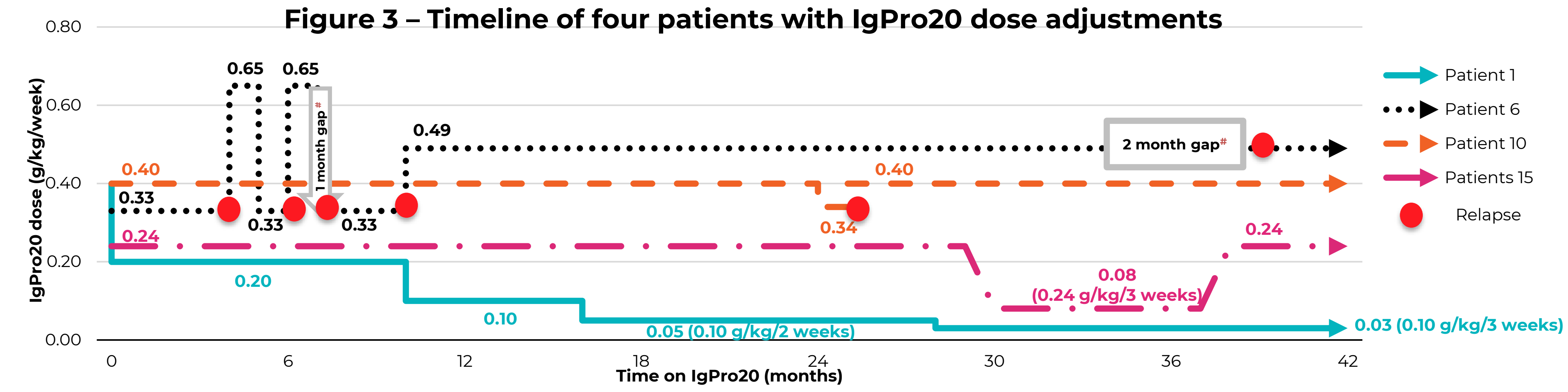


- Most physicians (n=16, 80.0%), shared information on the different treatment options for CIDP with their patients during the decision-making process

DOSE ADJUSTMENTS

- Ten patients (50.0%) had their IgPro20 dose adjusted at least once
  - Nine patients (45.0%) who were stable at the initial dose did not require any dose adjustments to maintain clinical stability, and no information was provided for one patient (5.0%)
- **Dose increases due to relapse of symptoms:** Five patients relapsed after 2-, 4-, 6-, 17-, and 42-months following transition to IgPro20, and required an increase in dose. One patient had their dose increased from 0.38 g/kg/week to 0.50 g/kg/week due to lack of improvement in symptoms
- **Dose decreases and frequency adjustments with improvement of symptoms:** Patient 1 (**Figure 3**) had their dose gradually tapered over 2.5 years. Following 2 years of clinical stability, Patient 10 (**Figure 3**) had their dose reduced, but the patient relapsed after 3 weeks and returned to their original dose. Patient 15 (**Figure 3**) was successfully maintained at lower doses during her pregnancy, when the frequency of her infusions was temporarily reduced
- **Case of multiple dose adjustments: Patient 6** (**Figure 3**) had their dose adjusted multiple times over the course of 3 years due to clinical instability at the lower dose and due to insurance coverage issues, resulting in temporary treatment gaps and subsequent relapse. Once these issues were resolved, the patient was clinically stable at 0.49 g/kg/week

Figure 3 – Timeline of four patients with IgPro20 dose adjustments



Insurance coverage issues caused a gap in receiving treatment

FOLLOWING DOSE ADJUSTMENT

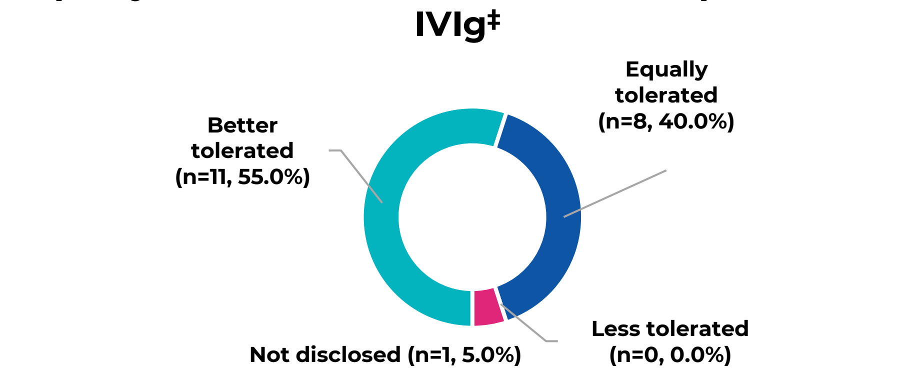
- Overall, the final mean (SD) IgPro20 dose was 0.32 (0.15) g/kg/week, with eight patients (40.0%) receiving an IgPro20 dose within the recommended dose range of 0.20–0.40 g/kg/week[2,5]
- No patients discontinued IgPro20 use due to adverse events† (The mean [SD] duration of IgPro20 use at the time of data collection was 4.2 [1.8] years [n=20])

PATIENT PERCEPTION

- Overall, 95% of patients felt IgPro20 was equally or better tolerated when compared with IVIg (**Figure 4**)

†The inclusion criteria of needing 6 months prior SCIg treatment may have eliminated patients who discontinued early to due adverse events

Figure 4 – Most patients perceived IgPro20 to be equally or better tolerated when compared with IVIg‡



‡The inclusion criteria of needing 6 months of prior SCIg treatment may have eliminated patients who perceived IgPro20 to be less tolerated compared with IVIg. IVIg, intravenous immunoglobulin; SCIg, subcutaneous immunoglobulin.

Discussion

- This study presents real-world evidence for the current use of IgPro20 in patients with CIDP in the US
- The case studies show that the 1:1 equivalent dose when transitioning from IVIg to SCIg, is the most commonly used dose
- Individualization of dosing is an important consideration for successful long-term treatment management
- Monitoring patient symptoms and involving the patient in treatment decisions is necessary to tailor dose adjustments for individual patient situations

Conclusions

- **The patient cases described here, demonstrate the clinical use of IgPro20 as maintenance therapy in CIDP, highlighting that individualization of dosing is an important consideration for long-term treatment management, that can have positive impacts on patient outcomes**
- **These results support evidence-based decision making by physicians in everyday clinical practice and the EAN/PNS recommendation for SCIg as a choice for maintenance therapy for CIDP**

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