Infusion Parameters and Demographics of Patients with Primary Immunodeficiency and Chronic Inflammatory Demyelinating Polyneuropathy During SCIg Self-administration Training

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Introduction

Primary Immunodeficiency (PI)

- PIs comprise more than ~350 different types of inherited diseases characterized by an impaired or absent components of the immune system[1]
  - Clinical features include increased susceptibility to infections and a higher risk of allergies, autoimmune diseases, and malignancy[2]

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- CIDP is a progressive immune-mediated disorder of the peripheral nerves attributed to demyelination and impaired signal conduction in motor and/or sensory nerves[3]
  - Clinical features include loss of sensation and progressive weakness

Treating PI and CIDP

- SCIg has been used for the long-term treatment of patients with PI for over a decade and was approved in March 2018 for maintenance therapy in adult patients with CIDP[4]
  - Compared with IVIg, the benefits of SCIg include self-administration at locations convenient for the patient, flexible dosing, and lower reported systemic reactions
- Patients are often trained to self-administer by specialty pharmacy nurses; the success of this training can affect whether the patient remains on SCIg or reverts back to their previous treatment

Objective

- **To analyze SCIg training data and outcomes collected by the Specialty Pharmacy Nurse Network (SPNN) in patients with PI and CIDP as they transitioned to SCIg**

Methods

SCIg Self-Administration Training

- This was a retrospective, observational study utilizing data collected by SPNN during the training of patients with PI or CIDP transitioning to SCIg between April 2017 and April 2019
  - Data collected included number of training sessions required; infusion parameters from each session; age, gender and body weight; and discontinuations and the reason for discontinuing
  - PI and CIDP study cohorts were identified via International Classification of Diseases (ICD) 10 codes: D80.0–84.9 for PIs and G61.81 for CIDP

Premier Start℠ Satisfaction Survey

- Upon completion of training, patients who had been assisted with the transition to SCIg by the Premier Start℠ Program were asked to complete a survey on their satisfaction with SCIg and confidence in self-administration
  - Data from responses collected between July 2018 and July 2019 were analyzed

Results

Patient Cohorts

- Of 1170 patients completing 1–7 SCIg training visits; 1001 were patients with PI and 169 were patients with CIDP
  - In the PI cohort, there was a 26%/74% male/female ratio and 140 (14%) were <18 years old
  - In the CIDP cohort, there was a 53%/47% male/female ratio and all were >18 years old

Results Continued

SCIg Training Requirements

- The majority of patients with PI (77%) required ≤3 training sessions before they were independent, whereas only 49% of patients with CIDP were independent with ≤3 training sessions
  - On average, patients with CIDP required 3.3 training sessions [range: 1–7]
  - However, infusion parameters were largely similar between patients at their final visit irrespective of how many training sessions they required (**Table 1**)
- Compared with their first visit, most patients with PI (65%) and CIDP (67%) were able to increase their infusion rate (mL/hr/site) by their final visit
- Mean infusion rates increased by 24% and 26% for PI and CIDP, respectively

Outcomes – Discontinuations

- In each cohort, **3.6% (n=36)** of patients with PI and **7.1% (n=12)** of patients with CIDP discontinued SCIg during training due to side effects
- Side effects were divided into systemic/local adverse events (AE) and worsening of symptoms (**Figure 1**)
- The most commonly-reported AEs in those who discontinued were:
  - Local site reactions (n=14) and fatigue (n=8) in the PI cohort
  - Fatigue (n=4) and nausea (n=4) in the CIDP cohort
- In the CIDP cohort, all 12 discontinuations were patients receiving 0.2 g/kg bodyweight (bw)
  - One of these was subsequently increased to 0.4 g/kg bw and then discontinued without adequate time for observation and dose adjustment

Local Site Reactions

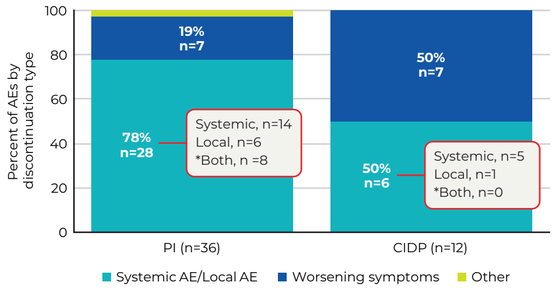
- In the PI cohort, there were 6 discontinuations as a result of intolerable local site reactions; reported local AEs were: swelling and redness at the infusion site; itchiness and rash at the infusion site; and persistent pain at the infusion site
- In the CIDP cohort, only 1 discontinuation was solely as result of an intolerable local site reaction; this was:
  - Swelling at the infusion site and lifestyle incompatibility

Table 1: Patient Characteristics and SCIg Infusion Parameters Stratifed by Training Requirements

	PI (N=1001)		CIDP (N=169)	
Training sessions required:	**≤ 3 (n=768)**	**> 3 (n=192)**	**≤ 3 (n=82)**	**> 3 (n=74)**
Demographics				
Mean age, years (SD)	48 (21.3)	54 (20.7)	55 (12.4)	59 (23.9)
Gender, % M/F	28/72	22/78	52/48	57/43
Mean BMI, kg/m² (SD)	28.0 (8.1)	25.6 (8.3)	30.0 (7.8)	30.7 (6.7)
Infusion Parameters*				
Mean infusion duration, mins (SD)	85.0 (36.4)	78.5 (26.9)	88.3 (35.6)	100.6 (25.7)
Mean number of sites (SD)	3.5 (1.2)	3.1 (1.0)	3.1 (1.1)	3.7 (1.1)
Mean dose, g (SD)	11.5 (5.4)	9.7 (4.1)	19.5 (8.4)	26.3 (9.4)
Mean infusion volume, mL (min, max)	54.6 (1, 200)	48.6 (10, 140)	97.7 (20, 270)	131.5 (40, 240)
Mean infusion rate/site, mL/hr/site (min, max)	13.3 (6.4, 37.2)	14.7 (6.1, 37.2)	23.7 (12.3, 32.2)	22.6 (13.1, 48.1)

Patients who did not complete training (discontinuations) not included. *As recorded at final training visit. BMI, body mass index; CIDP, chronic inflammatory demyelinating polyneuropathy; M/F, male/female ratio; PI, primary immunodeficiency; SD, standard deviation. SCIg, subcutaneous immunoglobulin.

Figure 1: Proportion of Discontinuations in Patients with PI and CIDP Due to Systemic/Local AEs, Worsening of Symptoms, or Other AE Reason*

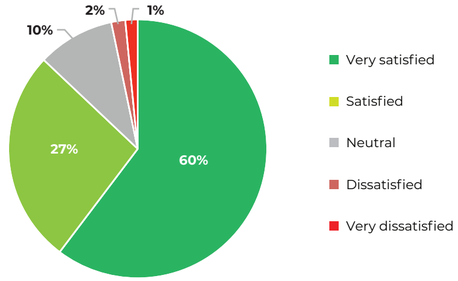


Other reason (3%), patient was diagnosed with lung cancer during training. AE, adverse event. *Gray boxes indicate breakdown of those reporting systemic, local, or both AEs.

Outcomes – Premier StartSM Satisfaction Survey Results

- Of 912 respondents, 794 (87%) were either satisfied or very satisfied with their confidence in their self administration ability following training (**Figure 2**)
  - Overall, 30 patients were dissatisfied or very dissatisfied, with 11 of these citing 'a complicated infusion process' as the main reason for their lack of confidence
- Of 899 respondents, 850 (95%) were either satisfied or very satisfied with the support provided during their SCIg training

Figure 2: Respondent Satisfaction with Confidence in Self-Administration Ability



Training Recommendations

- With training, patients can achieve an adequate level of ability, but this may take some longer than others to achieve
  - Nurse training techniques may need to differ depending on patient age, comorbidities, and SCIg volume requirements
  - Mastery of self-administration should include optimizing rate and volume per site so that patients are comfortable with ancillary supplies and assessment of site reactions in relation to technique and ancillary supplies

Discussion Points

- Training can typically be achieved within 4 weeks, however, management and optimizing both flow rates and volumes per site may require time beyond the training sessions
- Data presented here was captured during the training sessions (4 week period) and may not be reflective of the final infusion parameters that can be applied and achieved per the prescribing information and individual tolerability
- Maximizing flow rate can provide the patient with an infusion regimen that requires less time; maximizing volume per site can provide fewer needles sticks
- Additional consideration in regards to adjustment of ancillary supplies to mitigate local site reactions is important to maximize tolerability and increase compliance
- This short time frame limits scope regarding understanding worsening symptoms and management of dosing adjustment for CIDP
- In the PATH extension study; 90% of patients did not experience CIDP relapse on 0.4 g/kg compared with 48% receiving 0.2 g/kg[5]
  - Most patients (89%) who were up-titrated to 0.4 g/kg recovered within 4 weeks

Conclusions

- **Nurse confidence and experience with larger volumes is key to optimizing SCIg training and may result in patients' ability to learn self-administration more quickly and with fewer discontinuations**
- **Management of infusion parameters after training should continue until optimal infusion regimen can be achieved**
- **More data is needed to better explore the association between patient ability to infuse independently and the nursing techniques used for education**
- **The majority of patients (87%) with training assisted by the Premier Start℠ Program were satisfied or very satisfied with SCIg therapy and were confident self-infusing**

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Acknowledgments

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