Impact of CIDP on Daily Activity and Participation: I-RODS Analysis from a US Patient Survey

Ingemar S.J. Merkies,[1,2] Rajiv Mallick,[3] Anne Haudrich,[3] Ann L. Bullinger[3] and Catharina Faber[1]

[1]Department of Neurology, Maastricht University Medical Center, Maastricht, the Netherlands;
[2]Department of Neurology, St Elizabeth Hospital, Willemstad, Curaçao;
[3]CSL Behring, King of Prussia, PA, USA.

Introduction

- Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare peripheral neuropathy with an estimated prevalence of 1–9 cases per 100,000 individuals and an annual incidence of 0.5–1.6 per 100,000[1]
- The most common symptoms are difficulty walking and using arms and legs, muscle weakness and altered sensation (e.g., numbness and tingling)[2]

IMPACT IN REAL-WORLD SETTING

- Although treatable, following remission, CIDP is often associated with relapses during or after treatment, which can lead to permanent disability
- The impact on daily functional activity and participation in a real-world setting has previously gone undocumented

US PATIENT SURVEY

- Here, we employed a US nationwide survey of CIDP patients to assess the impact of disease-related disability to perform daily activities and assess the extent and effects of diagnostic delay

Aim

- To assess the impact of CIDP on disease-related daily activities and participation, and physical function

Methods

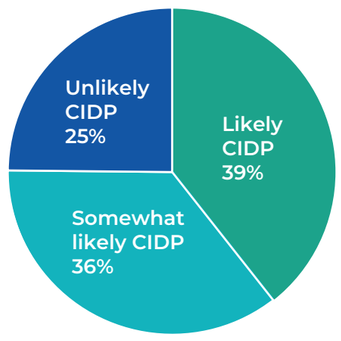
- Approximately 3250 individuals aged ≥18 years, recruited by the GBS|CIDP Foundation and self-reported to have CIDP, were invited to complete an online survey; of these, 475 completed the survey and their responses were used to assess daily activity and participation
- Patient diagnosis and treatment patterns were evaluated; impact on daily activity and participation was measured using the patient-reported Inflammatory Rasch-built Overall Disability Scale (I-RODS), and physical function was measured using the Patient Reported Outcomes Measurement Information System Physical Function T-score (PROMIS PF T-score)
- Data were analyzed overall and by stratification of patients based on the likelihood of an accurate CIDP diagnosis, defined as shown in **Table 1**

Table 1: Stratification of patients based on likelihood of accurate CIDP diagnosis

Unlikely CIDP patient
• Reported no muscle weakness as symptom of CIDP • Did not report having neurophysiologic tests performed when diagnosed
Somewhat likely CIDP patient
• Reported weakness, but not consistently • Reported symptoms were at their worst in less than 2 months (without prior diagnosis of Guillain-Barré syndrome [GBS]) • Reported symptoms were not symmetric
Likely CIDP patient
• Absence of the above-listed factors • Includes patients whose symptoms reached their worst in less than 2 months with a previous diagnosis of GBS

- There were 187, 170 and 118 patients with likely, somewhat likely and unlikely CIDP, respectively (**Figure 1**)

Figure 1: Percentage of respondents classified as either likely, somewhat likely or unlikely CIDP patients



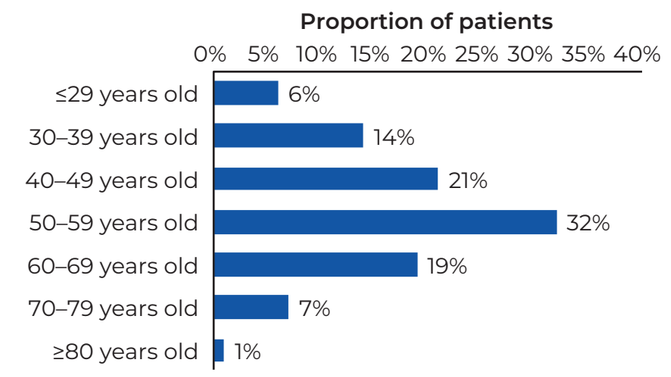
Patients were classified according to the criteria listed in Table 1.

Results

AGE AT CIDP DIAGNOSIS AND CURRENT TREATMENT

- The age distribution is shown in **Figure 2**; mean age at time of CIDP diagnosis was 51 years
- Common current treatments were intravenous immunoglobulin (63%), corticosteroids (19%) or immunosuppressive medicines (16%)

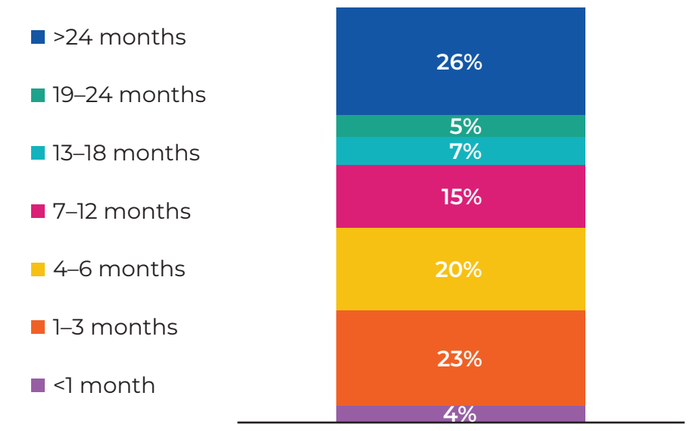
Figure 2: Age at the time of CIDP diagnosis



TIME BETWEEN FIRST SYMPTOMS AND DIAGNOSIS

- There was a median of 7 months between patients noticing the first symptoms and receiving their CIDP diagnosis (**Figure 3**)
  - For 26%, the time was more than 24 months

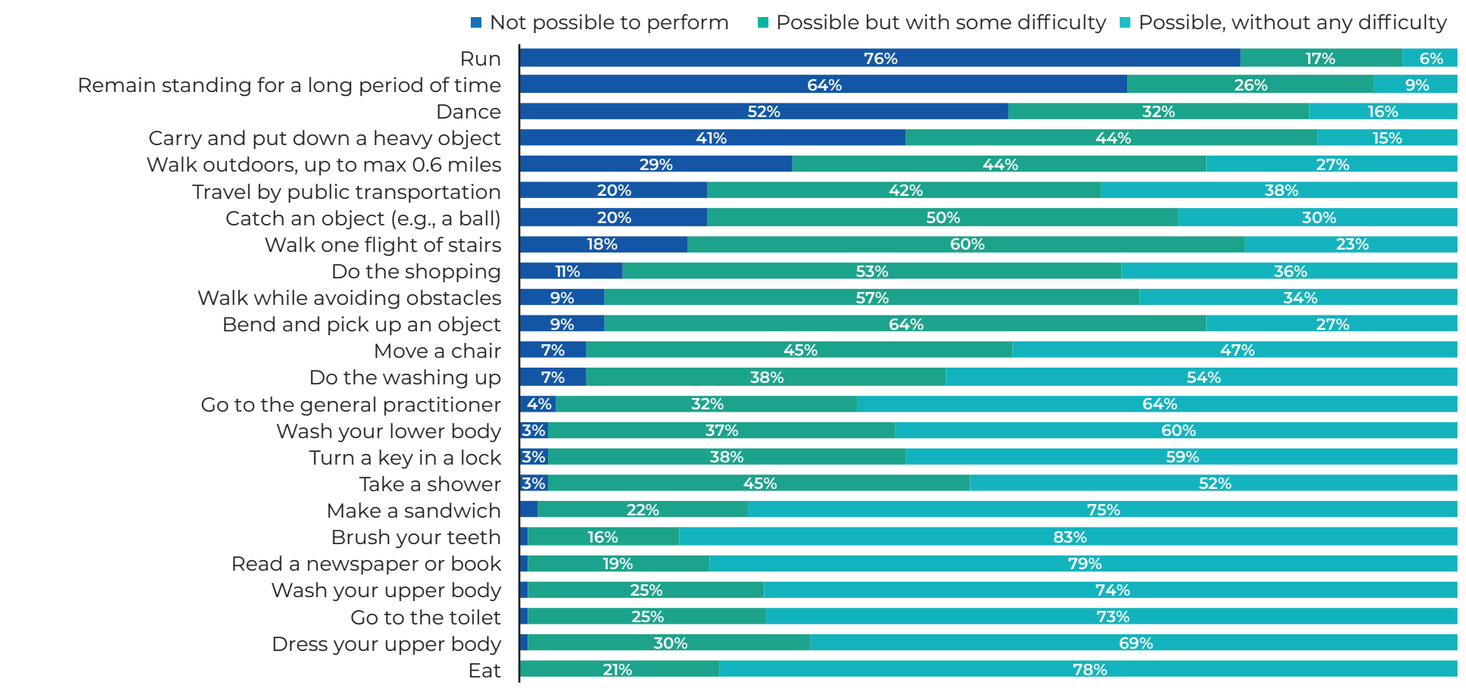
Figure 3: Time between noticing first symptoms and CIDP diagnosis



I-RODS: DIFFICULTY WITH PERFORMING DAILY ACTIVITIES

- A substantial subpopulation reported being unable to, or only with difficulty, perform the easiest items: going to the toilet (27%), eating (22%), reading a newspaper or book (21%) and brushing teeth (17%) (**Figure 4**)
- The majority of patients reported being unable to, or only with difficulty, perform activities at the middle-to-high difficultly level: walking outdoors (73%), walking one flight of stairs (77%) and running (94%) (**Figure 4**)

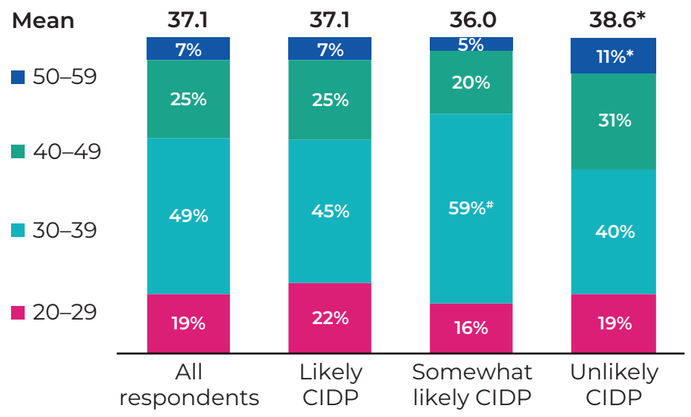
Figure 4: I-RODS: Proportion of patients experiencing difficulty with daily activities



PROMIS PHYSICAL FUNCTION T-SCORE: IMPACT OF LIKELIHOOD OF DIAGNOSIS

- Overall, 68% (49%+19%) of patients were more than one standard deviation [>10 points] below the US norm T-score of 50 (**Figure 5**)
  - Across strata, these percentages were 67% and 76% for those with likely or somewhat likely CIDP but 58% for "unlikely CIDP"
- Those with "unlikely CIDP" were least likely to be substantially impaired on physical function (T-score <40; p<0.05 vs "somewhat unlikely CIDP")

Figure 5: Impact of likelihood of CIDP diagnosis on PROMIS PF T-score

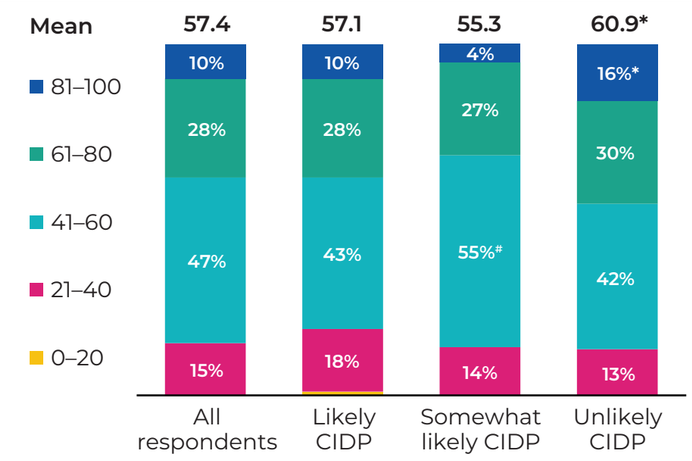


*p<0.05 vs somewhat likely CIDP; #p<0.05 likely and unlikely CIDP.

I-RODS: IMPACT OF LIKELIHOOD OF CIDP DIAGNOSIS

- Patients who were unlikely to have received an accurate CIDP diagnosis were most likely to have the best range of I-RODS centile scores (81–100) (**Figure 6**)

Figure 6: Impact of likelihood of CIDP diagnosis on I-RODS centile score



*p<0.05 vs somewhat likely CIDP; #p<0.05 vs likely and unlikely CIDP.

References

1. Dalakas MC. *Nat Rev Neurol*. 2011; **7**:507–517.
2. Mathey EK, et al. J Neurol Neurosurg Psychiatry. 2015; **86**:973–985.

Editorial assistance was provided by Meridian HealthComms.

Conclusion

- Findings from this nationwide US survey demonstrate that CIDP significantly impacts daily activity and participation, including the simplest daily activities
- Diagnosis of CIDP is delayed in a large number of patients

Presented at the 2018 Peripheral Nerve Society (PNS) Annual Meeting, Baltimore, MD, USA; July 22–25, 2018

CSL Behring