International Journal of Scientific Reports
Rajani A et al. Int J Sci Rep. 2022 Dec;8(12):362-365
http://www.sci-rep.com
pISSN 2454-2156 | eISSN 2454-2164
DOI: https://dx.doi.org/10.18203/issn.2454-2156.IntJSciRep20223001

Original Research Article

Disease profile of Guillain-Barré syndrome among Indian patients receiving intravenous immunoglobulin: a real-world observational study

Anil Rajani*, Lav Patel, Shreekant Sharma

Medical Affairs, Intas Pharmaceuticals Limited, Ahmedabad, Gujarat, India

Received: 20 October 2022
Revised: 01 November 2022
Accepted: 08 November 2022

***Correspondence:**
Dr. Anil Rajani,
E-mail: anil_rajani@intaspharma.com

ABSTRACT

Background: Objectives of the study were to evaluate the real-world usage pattern of intravenous immunoglobulin (IVIg) for the treatment of patients with Guillain-Barré syndrome (GBS), and to understand the disease characteristics and demographic patterns of these patients.
Methods: This real-world, retrospective, analysis included data of patients with GBS who received IVIg treatment at various centers across India. The study data was collected between April 2021 and March 2022.
Results: A total of 3064 patients with GBS who received IVIg treatment were included. The mean (SD) age of the patients was 48.97 (14.97) years, and majority of the patients were men (68.8%). Acute inflammatory demyelinating polyneuropathy (AIDP) was the most common subtype of GBS (60.5%), followed by acute motor axonal neuropathy (AMAN; 11.6%), and acute motor sensory axonal neuropathy (AMSAN; 3%). The majority (>94%) of patients received IVIg therapy as first-line treatment. A large proportion of the patients (n=2402, 78.4%) were given the standard dose of 2 g/kg bodyweight (given over 5 days) and 97.1% (n=2974) of the total study population received IVIg regimen over the standard protocol of 5 days.
Conclusions: In the clinical spectrum of GBS, AIDP was the most common subtype. IVIg was used at generally recommended dose and duration in patients with GBS.

Keywords: IVIg, Intravenous immunoglobulin, GBS, Guillain-Barré syndrome

INTRODUCTION

Guillain-Barré syndrome (GBS), an acute, immune-mediated paralysing, inflammatory peripheral nerve disease, is an uncommon disease of the nerves outside the brain and spinal cord.[1,2] It is characterized by weakness and numbness of the limbs, facial and respiratory muscles. The common pathogenesis includes multifocal inflammation of spinal roots and peripheral nerves, especially their myelin sheaths.[2-4]

GBS has a global median annual incidence of 1.11 (range: 0.81–1.89) cases per 100,000 persons.[5]

The GBS has been traditionally used interchangeably with acute inflammatory demyelinating polyradiculo-neuropathy (AIDP) for several years, until the recent evidence suggested that AIDP is a subtype of GBS among others. The GBS is considered as group of several immune polyneuropathies that leads to generalized weakness with different etiopathogenesis. Among these, the axonal subtypes of GBS include acute motor axonal neuropathy (AMAN) and acute motor-sensory axonal neuropathy (AMSAN). Furthermore, several other subtypes of GBS, have been reported.[6] The AIDP subtype of GBS is predominantly observed in Europe and North America whereas in Asian countries, AMAN is the most common subtype.[7]

Management with immunomodulatory drugs is recommended in GBS when patients are unable to walk independently for 10 metres.[8] In up to 30% patients with GBS diagnosis, mechanical ventilation and intensive care unit (ICU) admission are warranted.[9] Pharmacological treatments are effective in reducing the time to recovery and improving the clinical outcomes in patients with GBS.[10] Patients with GBS are commonly treated with intravenous immunoglobulins (IVIg), plasma exchange (PE) and experimental drugs such as eculizumab.[9,10]

The current study was conducted to evaluate the real-world usage patterns of IVIg for the treatment of patients with GBS, and to understand the disease characteristics and demographic pattern of patients with GBS receiving IVIg treatment.

METHODS

Study design

In this retrospective, observational study, data were collected from multiple centres across India including hospitals, clinics and health care institutes. The study inclusion criteria were the patients with GBS who received IVIg treatment at the study centers. Data were collected retrospectively by the neurologists in the GBS IVIg study data collection form. Data of patients with GBS, who received treatments other than IVIg were excluded. The choice of patient selection was purely based on treating physician's discretion. There were no new interventions or evaluations performed in this study.

The data was collected between April 2021 and March 2022. The data collected included demographic details (age, sex, height, weight etc), disease characteristics, medical history, details of treatment. The study endpoints included the descriptive analysis of patient demographic profile and disease characteristics, and the proportion of patients receiving various IVIg regimens.

Statistical analysis

There was no formal sample size calculation in this study, and the observations from patient's records only were analyzed. In this real-world study, patients' data was collected retrospectively. Demographic and baseline characteristics were summarized using descriptive statistics. Categorical variables were summarized with frequency and percentage. Continuous variables were summarized with count, mean, standard deviation, etc. Graphical presentation of data was done using bar chart as appropriate. Statistical analyses were performed using SAS® version 9.4 (SAS Institute Inc., USA).

Ethics statement

The study protocol was approved by the ACEAS independent ethics committee, Ahmedabad, India. This study was performed in accordance with Good Clinical Practice and the ethical principles of the Declaration of Helsinki. As this study involved data retrieval from patient records only, an informed consent of patients was not obtained.

RESULTS

Overall, data of 3064 patients with GBS who received IVIg treatment at 364 centres were analysed. The baseline characteristics of patients are summarized in Table 1. The mean age of the patients was 48.97 years. The gender distribution showed 68.8% men and 31.2% women in this study. Hypertension (41.1%) and diabetes (33.9%) were the most common comorbid conditions.

Table 1: Demographic profile.

Parameters	All patients (n=3064)
Age (years), mean±SD	48.97±14.97
Height in cm, mean±SD	162.57±13.82
Weight in kg, mean±SD	65.70±12.67
Gender, N (%)	
Men	2109 (68.8)
Women	955 (31.2)
Smoking status: yes, N (%)	816 (26.6)
History of alcohol intake: yes, N (%)	829 (27.1)
Comorbid conditions, N (%)	
Hypertension	1259 (41.1)
Diabetes	1040 (33.9)
Hepatic dysfunction	159 (5.2)
Renal dysfunction	160 (5.2)
Osteoporosis	155 (5.1)

Cm-centimetre, kg-kilogram, SD-standard deviation.

Disease characteristics

Bilateral and flaccid weakness was the most common presentation (65.3%) followed by decreased/absent reflexes (58.7%) (Figure 1).

Among antecedent conditions within the last 2 months, fever (37.2%) was most commonly reported followed by cough (28.5%) (Figure 2).

Subtypes of GBS

As per electrophysiological studies, acute inflammatory demyelinating polyneuropathy (AIDP) was found to be the most common subtype of GBS (60.5%), followed by acute motor axonal neuropathy (AMAN; 11.6%), and acute motor sensory axonal neuropathy (AMSAN; 3%) (Table 2).

Details of the therapy

The majority (>94%) of patients received IVIg therapy as first-line treatment (Table 3). Among patients receiving

IVIg as later line of therapy, steroids were generally given in their earlier lines of therapy and very few patients had undergone plasmapheresis. A total of 2402 (78.4%) patients were given the standard dose of 2 g/kg bodyweight (given over 5 days); and 97.1% (n=2974) of the total study population received IVIg regimen over 5 days as per the standard protocol.

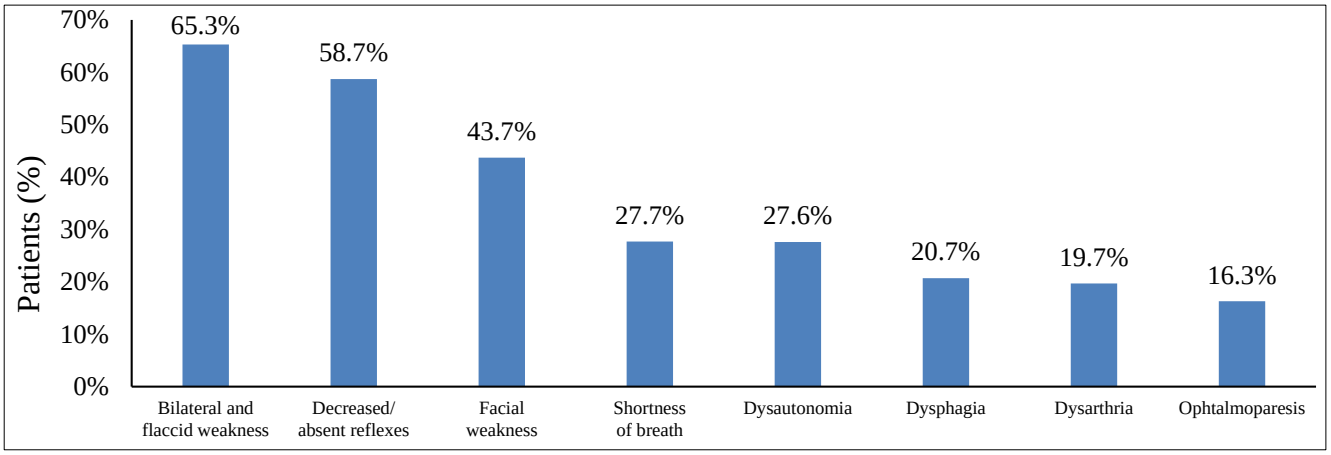

Figure 1: Clinical presentation.

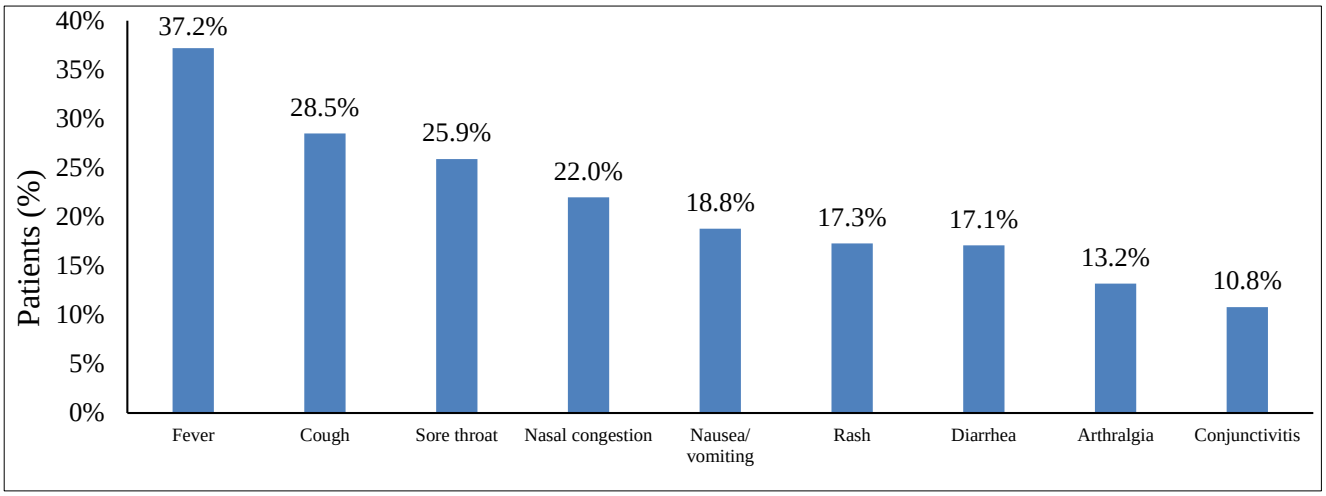

Figure 2: Antecedent conditions within the last 2 months.

Table 2: Subtypes of GBS as per electrophysiological studies.

Type of GBS, N (%)	All patients (n=3064)
AIDP	1855 (60.5)
AMAN	356 (11.6)
AMSAN	91 (3.0)
Equivocal or inexcitable	762 (24.9)

AIDP-acute inflammatory demyelinating polyneuropathy; AMAN-acute motor axonal neuropathy; AMSAN-acute motor and sensory axonal neuropathy.

Table 3: Details of the therapy.

Line of therapy, N (%)	All patients (n=3064)
1st	2908 (94.9)
2nd	151 (4.9)
3rd	5 (0.2)

DISCUSSION

This retrospective study in 3064 patients with GBS showed that the real-world usage pattern of IVIg was largely aligned with the published literature. AIDP was the most common subtype of GBS. The dosing and duration of IVIg were in line with the recommended treatment protocol.

In this study, the mean age of the patients was 48.97 years, with a male preponderance (male: female, 2.2:1). Dhadke et al reported similar findings with a male preponderance (male: female, 1.5:1) and commonest age group affected was 13-40 years in a real-world study.[11] Rath et al reported a male: female ratio of 1.5:1 and that median age of 48 years in GBS patients in a real-world study.[12]

In our study, bilateral and flaccid weakness was the most common presentation followed by decreased/absent

reflexes, which is generally consistent with previous reports.[1,8] Fever and cough were the most common antecedent conditions within the last 2 months in our study, similar to that observed by Koga et al.[13]

Our study reported that AIDP (60.5%) was the most common subtype of GBS followed by AMAN (11.6%) and AMSAN (3%) types. In India, Sriwastava et al reported that AIDP was the most common GBS subtype reported in 66% of the patients; AMSAN (4%) and AMAN (2%) were also reported.[14] Kalita et al reported that 73.8% patients had AIDP, 13.4% had AMAN, and 4.6% had AMSAN subtypes.[15]

IVIg is a proven effective treatment for GBS. The exact mechanism of IVIg in GBS remains unclear, however, it is thought to have immune-modulating action. Rath et al reported that IVIg was given over 5 days in majority of the courses (81%) in GBS patients.[12] In our study majority of the patients received the standard dose of 0.4 g/kg body weight daily for 5 days, and mostly patients were on 5-day regimen. The current study findings interpretation require consideration in view of certain limitations which include missing data, and potential inconsistency in data entry as multiple study centres were involved. Future prospective studies are warranted to elicit efficacy and safety results of IVIg in patients with GBS.

CONCLUSION

This retrospective, observational study reports the real-world usage patterns of IVIg in GBS patients in India. In the clinical spectrum of GBS, AIDP was the most common subtype. IVIg was used at generally recommended dose and duration in Indian patients with GBS.

ACKNOWLEDGEMENTS

Authors would like to thank the study investigators for their contribution.

Funding: The study was funded by Intas Pharmaceuticals Limited
Conflict of interest: Dr. Anil Rajani, Dr. Lav Patel, and Mr. Shreekant Sharma are employees of Intas Pharmaceuticals Limited, Ahmedabad, Gujarat, India
Ethical approval: The study was approved by the ACEAS Independent Ethics Committee, Ahmedabad, India

REFERENCES

1. Nguyen TP, Taylor RS. Guillain Barre Syndrome. [Updated 2022 Jul 4]. In: StatPearls. Treasure Island (FL): StatPearls Publishing. 2022.
2. Van den Berg B, Walgaard C, Drenthen J, Fokke C, Jacobs BC, van Doorn PA. Guillain–Barré syndrome: pathogenesis, diagnosis, treatment and prognosis. Nat Rev Neurol. 2014;10:469-82.
3. Van Doorn PA, Kuitwaard K, Walgaard C, van Koningsveld R, Ruts L, Jacobs BC. IVIG treatment and prognosis in Guillain-Barré syndrome. J Clin Immunol. 2010;30(1):S74-8.
4. Hughes RA, Swan AV, van Doorn PA. Intravenous immunoglobulin for Guillain-Barré syndrome. Cochrane Database Syst Rev. 2014;Cd002063.
5. Benedetti L, Briani C, Beronio A, Massa F, Giorli E, Sani C, et al. Increased incidence of axonal Guillain-Barré syndrome in La Spezia area of Italy: A 13-year follow-up study. J Peripher Nerv Syst. 2019;24:80-6.
6. Piccione EA, Salame K, Katirji B. Guillain-Barré Syndrome and Related Disorders, in Neuromuscular Disorders in Clinical Practice. Katirji B, Kaminski HJ, Ruff RL. Springer New York: New York, NY. 2014;573-603.
7. Meena AK, Khadilkar SV, Murthy JM. Treatment guidelines for Guillain-Barré Syndrome. Ann Indian Acad Neurol. 2011;14:S73-81.
8. Leonhard SE, Mandarakas MR, Gondim FAA, Bateman K, Ferreira MLB, Cornblath DR, et al. Diagnosis and management of Guillain-Barré syndrome in ten steps. Nat Rev Neurol. 2019;15:671-83.
9. Shang P, Feng J, Wu W, Zhang H-L. Intensive Care and Treatment of Severe Guillain–Barré Syndrome. Front Pharmacol. 2021;12.
10. Rath J, Zulehner G, Schober B, Grisold A, Krenn M, Cetin H, et al. Real-world treatment of adult patients with Guillain-Barré syndrome over the last two decades. Scientif Rep. 2021;11:19170.
11. Dhadke SV, Dhadke VN, Bangar SS, Korade MB. Clinical profile of Guillain Barre syndrome. J Assoc Physicians India. 2013;61:168-72.
12. Rath J, Zulehner G, Schober B, Grisold A, Krenn M, Cetin H, et al. Real-world treatment of adult patients with Guillain-Barré syndrome over the last two decades. Sci Rep. 2021;11:19170.
13. Koga M, Yuki N, Hirata K. Antecedent symptoms in Guillain-Barré syndrome: an important indicator for clinical and serological subgroups. Acta Neurol Scand. 2001;103:278-87.
14. Sriwastava S, Kataria S, Tandon M, Patel J, Patel R, Jowkar A, et al. Guillain Barré Syndrome and its variants as a manifestation of COVID-19: A systematic review of case reports and case series. J Neurol Sci. 2021;420:117263.
15. Kalita J, Misra UK, Goyal G, Das M. Guillain-Barré syndrome: subtypes and predictors of outcome from India. J Peripher Nerv Syst. 2014;19:36-43.

Cite this article as: Rajani A, Patel L, Sharma S. Disease profile of Guillain-Barré syndrome among Indian patients receiving intravenous immunoglobulin: a real-world observational study. Int J Sci Rep 2022;8(12):362-5.