

Guillain-Barre Syndrome, Clinical Spectrum and Electrophysiological Sub Types: A Local Experience

Faryal Shamim¹, Safia Bano², Masooma Waqar³, Ahsan Numan⁴

Department of Neurology, King Edward Medical University, Lahore Pakistan^{1,2,3,4}

ABSTRACT

Background: Guillain-Barre syndrome is an uncommon neurological disease that occurs in the acute phase as generalized polyradiculo-neuropathy. It has multiple variants some of them being uncommon and rarely encountered.

Objective: The objective of the study was to determine the frequency and clinical characteristics of Guillain-Barre syndrome variants.

Methods: A cross-sectional study was carried out in the neurology department, at Mayo hospital from July to December 2022. Patients clinically diagnosed as Guillain-Barre syndrome by using Brighton criteria were included. All the patients underwent electrodiagnostic procedures and they were categorized into five basic variants i.e., Acute motor axonal neuropathy, Acute motor sensory axonal neuropathy, acute inflammatory demyelinating polyradiculo-neuropathy, acute sensory axonal neuropathy and mixed variety acute polyradiculo-neuropathy.

Results: Out of total 85 patients, males were 56 (65.9%) and 29 (34.1%) were females. Adults were more affected with a mean age of 36±15.3. The frequency of acute motor axonal neuropathy (35.3%) was high followed by acute motor sensory axonal neuropathy (31.8%), mixed variety acute polyradiculo-neuropathy (16.5%), acute inflammatory demyelinating polyradiculo-neuropathy (12.9%) and acute sensory axonal neuropathy (3.5%) respectively.

Conclusion: Acute motor axonal neuropathy is frequently encountered Guillain-Barre syndrome variant in our setup. Also, the frequency of acute sensory axonal neuropathy and mixed variety acute polyradiculo-neuropathy has increased with a declining trend of acute inflammatory demyelinating polyradiculo-neuropathy.

Key Words: Guillain-Barre syndrome, acute motor axonal neuropathy, acute motor sensory axonal neuropathy, acute inflammatory demyelinating polyradiculo-neuropathy, acute sensory axonal neuropathy, Electro diagnostic

Corresponding Author:

Dr Safia Bano
Assistant Professor
Department of Neurology
King Edward Medical University, Lahore
Pakistan

Email address: safiabano207@gmail.com

Received: 23.02.2023, Revised: 12.06.2023,

Accepted: 23.06.2023

How to cite this article: Shamim F, Bano S, Waqar M, Numan A. Guillain-Barre Syndrome, clinical spectrum and electrophysiological subtypes: A Local experience. J Shalamar Med Dent Coll. 2023, 4(1): 60-65. doi: 10.53685/jshmdc.v4i1.150

Doi: <https://doi.org/10.53685/jshmdc.v4i1.150>



This is an open-access article distributed under the terms of the Creative Commons Attribution-Noncommercial 4.0 International license.

INTRODUCTION

Guillain-Barre syndrome (GBS) is a post-infectious disorder that presents as acute polyradiculo-neuropathy that may be accompanied by acute flaccid paralysis or rapidly ascending body weakness and loss of

sensations.¹ Symptoms and clinical presentation may vary depending upon the type of variant. GBS is often post-infectious or immune-mediated.² Temporal course for GBS is from days to weeks with the onset to nadir at 7-15 days, usually within 2 weeks.³ GBS often lasts for 3-4 weeks. Progression of disease beyond 1 month is rare in GBS patients.^{2,4}

Previously acute inflammatory demyelinating polyradiculo-neuropathy (AIDP) was a frequently encountered variety but now the prevalence of acute motor axonal neuropathy (AMAN) is increasing in South Asian countries. Moreover, the rare type of GBS i.e., acute sensory axonal neuropathy (ASAN) and mixed variety acute polyradiculo-neuropathy have an ascending number of patients encountered over recent years.⁵

Electro diagnostic (EDX) studies particularly nerve conduction studies play a very important diagnostic role in patients with GBS. EDX plays a crucial role in categorized the disease, its early detection and treatment of GBS. Diagnostic yield increases if nerve conduction studies are performed between 7-15 days of onset. This is a part of the standard workup for GBS. Patients respond well to intravenous gamma globulin (IVIG) or plasmapheresis therapy.⁶

Previous studies prove that AIDP is frequently seen variety of GBS in the Middle East but in South Asia AMAN is a more common finding with AMAN 32.78%, AMSAN 28.33% and AIDP 31.11%.⁷

The purpose of the study was to determine the clinical spectrum, its clinical and electrophysiological variants of our population because each variant has its clinical characteristics and prognosis. It will help clinicians to diagnose disease early and can offer prompt treatment. It will also help physicians to understand global versus regional epidemiological differences in GBS variants.

METHODS

A cross-sectional study was carried out in the

neurology department, of Mayo Hospital, Lahore. Permission was taken by the ethical committee (No.794/RC/KEMU) of King Edward Medical University before conducting the research. Patients presenting with symptoms of GBS in the outpatient department, emergency department and neurology ward, were included in the study from July 2022 to December 2022.

Nerve conduction studies were carried out on 85 consecutive cases who were clinically diagnosed as GBS according to Brighton criteria³ (i.e. history of weakness of limbs with or without sensory disturbance, involvement of the cranial nerve, electrophysiological findings, 'presence of delayed latency, reduced amplitude, slowed conduction velocity, delayed F-waves in tested nerves).

Patients with a history and examination suggestive of periodic paralysis, polyneuropathy other than GBS were excluded from study. Nerve conduction studies were done according to the EDX protocol for GBS between 7-15 days from the onset of symptoms in all the patients.

All demographic variables along with clinical spectrums and electrophysiological variants of GBS² (i.e. on the basis of involvement of either sensory and motor nerves or both on EDX determined by the presence of delayed latencies, reduced amplitude, slowed conduction velocity, delayed F-waves in tested nerves as compared with reference values) were recorded on predesigned performa.

Statistical Analysis

The data was analyzed using SPSS version 26. Frequencies and percentages were calculated for categorical/qualitative data i.e. gender, clinical severity, and GBS variants. Mean and standard deviation were calculated for numerical/quantitative data.

RESULT

85 patients from 15-70 years of age were selected and proceeded upon for the study. Out of which males were 56 (65.9%) and 29(34.1%) females. Adults were more affected with a mean± SD age of 36 ± 15.3.

Table 1: Demographics and Disease Characteristics	
Distribution in age groups	
Age group (years)	n (%)
11-20	17(20.0)
21-30	21(24.7)
31-40	11(12.9)
41-50	18(21.1)
51-60	14(16.4)
61-70	04(4.7)
Gender	n (%)
Males	56(65.9)
Females	29(34.1)
Symmetric flaccid limb Weakness	
	n (%)
Weakness of all four limbs (0/5 power)	28(32.9)
Weakness of all four limbs(3/5 power)	31(36.5)
Weakness of lower limbs	23(27.1)
No weakness with loss of sensations	03(3.5)
CSF Proteins	n (%)
Normal	16(18.8)
High	69(81.2)
Reflexes	n (%)
Normal	16(18.8)
Diminished	69(81.2)

The degree of limb weakness was as follows i.e., weakness of all four limbs with power=0/5 was seen in 32.9%, weakness of all four limbs (power=3/5) in 36.5%, generalized weakness of lower limbs only in 27.1% and no weakness with loss of sensation 3.5%. CSF proteins were high in 81.2% of patients and normal in 18.8% patients only. Other demographic and disease characteristics are given table 1. Frequently encountered variant of GBS was AMAN n=30 (35.3%) followed by AMSAN n=27 (31.8%), mixed variety acute polyradiculo-neuropathy n=14 (16.5%), AIDP n=14 (12.9%) and n=03 ASAN (3.5%) respectively.

Table 2: Frequency of Electrophysiological variants of GBS	
	n (%)
Acute motor axonal neuropathy	30(35.3)
Acute motor sensory axonal neuropathy	27(31.8)
Acute inflammatory demyelinating neuropathy	11(12.9)
Mixed variety of acute polyneuropathy	14(16.5)
Acute sensory Neuropathy	03(3.5)

DISCUSSION

GBS is a post-infectious disorder of peripheral nerves. The immune system mistakenly attacks nerves often preceding an upper respiratory tract infection.²

Most studies done in the past suggest AIDP being most common variant. A study done in Iran in 2014 by Yadegari S et al. states that AIDP had highest prevalence of 63%, AMAN 23%, AMSAN 14% only⁸ A research done in Pakistan in 2006 Zaheer et al. reveals that AIDP had a prevalence of 36% while AMAN 12% that is different from our study results.⁹ Another study shows a relatively close results with AIDP 46% and axonal variants 31% collectively.¹⁰

Whereas, in China research by Tian J et al. in 2019 showed that AMAN was common prevalent variety of GBS being 55.8% , AIDP 21.2%.¹¹ The results are close to our research results. Recent research in Pakistan also states axonal variants to be most prevalent being 61.1% in which AMAN contributed 32.78% and AMSAN contributed 28.3%. Prevalence of AIDP was found to be 31.1% being second most common variant that is contrary to our research.^{7,12}

Research in north India reveals that AMAN was 69.4 and AIDP was 25% prevalent in children with GBS.¹² Japan documents the highest prevalence of AMAN 45-48%.¹³ Another Chinese study reveals that AMAN was 55% prevalent while AIDP was 32% .¹⁴

The present study revealed the significant number of some rare variants i.e. mixed variety acute polyradiculo-neuropathy (16.5%), acute sensory axonal neuropathy (3.5%). literature has sparse data regarding these variants, and only a few case reports are available.¹⁵

Differences of geographic zones contribute to differences in the prevalence of GBS varieties even in the prevalence of classical GBS. The prevalence is least in Europe and North America which contribute only 5% to the total cases of GBS as compared to Asia^{15, 16}

Also, AIDP is still the one with the highest incidence in these regions.

A major alarming situation is the increase in prevalence of mixed variety acute polyradiculoneuropathy and acute sensory axonal variants. The cause of change in the pattern of prevalence of different subtypes is not clearly known yet but a study attributed it to *Campylobacter Jejuni* bacteria causing higher prevalence of AMAN.^{17,18} Due to lack of research and awareness on pure sensory axonal variant it is often clinically neglected.¹⁹ Our study revealed that every 3.5 out of 100 patients of GBS fall in the pure sensory axonal variant (ASAN) and every 16.5 out of 100 GBS patients fall in mixed variety variant category.

Similar to many other studies done in the past GBS is more common in males as compared to females.^{7,19,20} This may be attributed to the fact that males have a higher exposure to the environment thus viruses and bacteria. GBS is more common in our setup as compared to the western world is due to unhygienic environments, lack of proper sanitation in public places, and lack of awareness in the masses. Another cause for the higher prevalence of GBS is antibiotics misuse that leads to antibiotic resistant thus not able to resist infections leading to serious complications.

A study conducted by Siddiqui, Maimoona, et al, in Pakistan, revealed AIDP(52%) is more prevalent than AMAN(29%), the results are different from our study as AMAN variety is more common. This might be due to patient's regional difference (presenting from northern areas of Pakistan).²⁰ Our results regarding AMAN as common variant are consistent with study conducted in Khyber Pakhtunkhwa.²¹

Study limitation is, it is carried out in single center i.e., no patients other than Mayo hospital were sampled.

Secondly, prognosis and outcomes were not studied as follow-ups were not held. The cause of the disease was also not studied at the time of presentation.

CONCLUSION

Axonal variants of GBS is becoming more common over the years. The spectrum of subtypes is changing i.e., AMAN is increasing over time with a decline in the prevalence of AIDP. New variants of GBS are frequently encounter in clinical practice i.e., ASAN and mixed variety acute polyradiculo-neuropathy. Such variants should be suspected in atypical presentation of GBS.

This will prevent delay in diagnosis and treatment resulting in the reduction of disease-related morbidity and mortality. Future research should be done in order to find the reason of these rare variants.

Conflict of Interest:

The authors declared no conflict of interest

Contributors:

FS: Writing original draft, review and editing

SB: Conceptualization and methodology

MW: Data collection, data analysis

AN: Methodology, critical review.

All authors approved the final version and signed the agreement to be accountable for all aspects of the work.

Grant Support and Financial Disclosure:

No specific grant was taken for this research from any funding agency in the public, commercial or not-for-profit sectors.

Data Sharing Statement:

The data that support the findings of this study are available from the corresponding author on reasonable request.

REFERENCES

1. Dimachkie MM, Barohn RJ. Guillain-Barré syndrome and variants. *Neurol Clin.* 2013; 31(2): 491–510.doi:10.1016/j.ncl.2013.01.005
2. Giacomini PS. Electromyography and Neuromuscular Disorders: Clinical Electrophysiologic Correlations. *Mcgill J Med.* 2006; 9(2):173.
3. Fokke C, Van den Berg B, Drenthen J, Walgaard C, Van Doorn PA, Jacobs BC.

- Diagnosis of Guillain-Barré syndrome and validation of brighton criteria. *Brain*. 2014; 137(1): 33-43.doi:10.1093/brain/awt285
4. Bölükbaşı F, Ersen G, Gündüz A, Karaali-Savrun F, Yazici S, Uzun N, et al. Guillain-Barré Syndrome and Its Variants: Clinical Course and Prognostic Factors. *Noro Psikiyatı Ars*. 2019; 56(1):71-74.doi:10.5152/npa.2017.18091.
 5. Levin KH. Variants and mimics of Guillain Barré syndrome. *Variants and mimics of Guillain Barré syndrome*. *The Neurologist*. 2004; 10(2): 61-74.doi:10.1097/01.nrl.0000117821.35196.0b
 6. Arends S, Drenthen J, Van den Bergh P, Franssen H, Hadden RDM, Islam B, et al. Electrodiagnosis of Guillain-Barre syndrome in the International GBS Outcome Study: Differences in methods and reference values. *Clin Neurophysiol*. 2022; 138:231-240.doi: 10.1016/j.clinph.2021.12.014.
 7. Khan MW, Hussain A, Zeeshan HM. Electrophysiological Variants of Guillain Barre Syndrome (GBS). *Med.Forum*. 2020; 31(12): 20208.
 8. Yadegari S, Nafissi S, Kazemi NJIjon. Comparison of electrophysiological findings in axonal and demyelinating Guillain-Barre syndrome. *Iran J Neurol*. 2014; 13(3):138-143.
 9. Zaheer M, Naeem M, Nasrullah M. Electrophysiological pattern of neuropathy in Guillain-Barre syndrome. *Ann King Edward Med coll*.2006;12(4).doi:10.21649/akemu.v12i4.961
 10. Shafqat S, Khealani BA, Awan F, Abedin SE. Guillain-Barre syndrome in Pakistan: similarity of demyelinating and axonal variants. *Eur J Neurol*. 2006; 13(6):662–665. doi:10.1111/j.1468-1331.2006.01071.x
 11. Tian J, Liu X, Zhang K, Cao C, Li T, Li P, et al. Electrophysiological subtypes and prognostic factors of Guillain-Barre Syndrome in northern China. *Frontiers in Neurol*. 2019; 10:714.doi:10.3389/fneur.2019.00714
 12. Yadav S, Jain P, Sharma S, Kumar V, Aneja S. Guillain–Barre syndrome in North Indian children: Clinical and serial electrophysiological features. *Neurol Ind* 2019; 67: 724-727. doi:10.4103/0028-3886.263191
 13. Hughes RA, Cornblath DR. Guillain-barre syndrome. *The Lancet* 2005; 366(9497): 1653-1666.doi:10.1016/S0140-6736(05)67665-9
 14. Ho TW, Mishu B, Li CN, et al. Guillain Barre syndrome in Northern China. Relationship to *Campylobacter jejuni* infection and anti-glycolipid antibodies. *Brain* 1995; 118:597-605.doi:10.1093/brain/118.3.597
 15. Bano S, Sardar Z, Ahmar M, Liaquat S, Shafiq B, Numan A. Clinical spectrum and outcome of guillain-barré syndrome with plasmapheresis. *Indian J Med Spec*. 2022; 13(4): 226.doi:10.4103/Injms.injms_50_2
 16. Nagasawa K, Kuwabara S, Misawa S, Fujii K, Tanabe Y, Yuki N, et al. Electrophysiological subtypes and prognosis of childhood Guillain Barre syndrome in Japan. *Muscle Nerve* 2006; 33(6):766–770. doi:10.1002/mus.20520
 17. Devos D, Magot A, Perrier-Boeswillwald J, Fayet G, Leclair-Visonneau L, Ollivier Y, et al. Guillain-Barré syndrome during childhood: Particular clinical and electrophysiological features. *Muscle Nerve* 2013; 48: 247-251. doi: 10.1002/mus.23749
 18. Bae JS, Yuki N, Kuwabara S, Kim JK, Vucic S, Lin CS, Kiernan MC. Guillain–barré syndrome in asia. *J. Neurol. Neurosurg. Psychiatry*. 2014; 85(8):907-913.doi:10.1136/jnnp-2013-306212
 19. Kofahi R, Aldabbour B, Aljezawi ME. A Rare Case with New Insights: Pure Sensory Guillain Barre Syndrome with Axonal Features. *Int Med Case Rep J*. 2020; 543-549. doi:10.2147/IMC.RJ.S280255
 20. Siddiqui M, Majid S, Yusuf H, Mateen F. Electrophysiological Pattern and Predictors of Functional Outcome of Patients with

Guillain Barre Syndrome at a Tertiary Care Hospital in Pakistan. *J Coll Physicians Surg Pak.* 2022; 32(3):364-368.doi:10.29271/jcps p.2022.03.364

21. Ayaz ul Haq M, Nabi D, Khan MO, Ullah R, Junaid M, Nasarullah HM. Frequency,

age, gender distribution, and seasonal variation of Guillain-Barré syndrome in a Province of Pakistan: A Retrospective Study: Prevalence of Guillain-Barre Syndrome. *PJMHS.* 2023: 207-210.doi:10.54393/pjhs.v4i03.565