Original Article

Electrophysiological Variants of Guillain Barre Syndrome (GBS)

Variants of Guillain Barre Syndrome

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ABSTRACT

Objective: This study aims at identifying frequency and distribution of common electrophysiological variants according to age and gender in our region.

Study Design: A Cross-sectional study

Place and Duration of Study: This study was conducted at the department of Neurology Sheikh Zayed Medical College/Hospital, Rahim Yar Khan from Jan 2015 to October 2020.

Materials and Methods: Patients clinically and electro diagnostically diagnosed as GBS in the last five years were included in the study. After using the electro diagnostic criteria, patients were categorized into three main variants: AMSAN (acute motor-sensory axonal neuropathy), AMAN (acute motor axonal neuropathy), and AIDP (acute inflammatory demyelinating polyneuropathy).

Results: 180 patients included in the study of which 62.22% were male and 37.78% were females. About 28.33% had AMSAN, 32.78% had AMAN, and 31.11% had AIDP, respectively. Males are affected more in each type. There was not much difference in the incidence of different variants in our region.

Conclusion: Different variants of GBS occur with slight variation in incidence depending upon the criteria used to classify as axonal or demyelinating variety.

Key Words: Guillain Barre Syndrome (GBS), Acute Motor Sensory Axonal Neuropathy (AMSAN), Acute Motor Axonal Neuropathy (AMAN), Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), Nerve Conduction Studies (NCS).

Citation of article: Khan MWA, Hussain A, Zeeshan HM. Electrophysiological Variants of Guillain Barre Syndrome (GBS). Med Forum 2020;31(12):8-11.

INTRODUCTION

Guillain-Barre syndrome (GBS) is an immune mediated acute, generalized paralytic polyradiculoneuropathy that is characterized by progressive weakness and diminished or absent deep tendon reflexes with intact sphincter control and without a sensory level.[1,2]

In about two thirds of cases, GBS is preceded by a symptomatic infection such as Campylobacter jejuni, Epstein-Barr virus, influenza, Mycoplasma pneumonia, Haemophilus influenza or cytomegalovirus.[3] There has also been associations with influenza infections and vaccinations.[4] GBS cases were also reported during the Zika virus outbreak in South America.[5]

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Received: October, 2020
Accepted: November, 2020
Printed: December, 2020

GBS associated with COVID-19 is also reported, but the strength and mechanism of association remains unclear.[6]

Guillain-Barre syndrome (GBS) usually presents sporadically with global incidence reported around 0.6-2.4 cases per 100,000 populations. Men are affected by approximately 1.5 times than women.[7]

Apart from clinical and laboratory findings, electro diagnostic studies (EDX) are helpful in classification of different subtypes of the disease. Electro physiologically, GBS has three major subtypes: acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), and acute motor sensory axonal neuropathy (AMSAN). Predominant subtype of GBS differs according to the geographic area.[2]

AIDP is the most commonly occurring subtype in North America and Europe, about 90 % of all cases. However, in Asia, Central and South America axonal variants of GBS i.e. acute motor axonopathy (AMAN) and acute motor sensory axonopathy (AMSAN) are found to represent 30% to 47% of cases.[8,9,14]

Nerve Conduction Studies (NCS) can be recorded by the end of the first week of illness and are most pronounced by the second week. The diagnostic yield of NCS can be increased by recording at least four motor nerves, three sensory nerves, F waves and H-reflexes.[10]

The EDX findings supportive of AIDP include, prolonged distal motor latencies, reduced conduction velocities, conduction blocks at non-entrapment sites, temporal dispersion and prolonged F wave latencies. Another characteristic electro diagnostic feature of GBS is "sural sparing".[11] A normal sural sensory nerve action potential (SNAP) with abnormal upper extremity sensory nerve responses. This is very unlikely for other neuropathies than AIDP to manifest. Sural sparing, persists even in the later part of the disease. NCS in GBS is also helpful in assessing the need and duration of ventilatory support. Low compound muscle action potentials (CMAPs) are most predictive of a poor prognosis.[12, 13]

The studies on electrophysiological patterns and variants of GBS in Pakistan have shown a trend similar to other Asian countries.[9,14] However, there is no study done in the South Punjab region to identify the variants of GBS. The purpose of this study is to identify the electrophysiological variants of GBS and find out the most common type in our region.

MATERIALS AND METHODS

This is an analytical cross sectional study conducted at the Department of Neurology Sheikh Zayed Medical College/Hospital Rahim Yar Khan. The study included patients who were diagnosed clinically and confirmed electro diagnostically to have GB syndrome. The data was collected from the last five years from January 2015 to October 2020. The patients were admitted from ER, OPD and Medical Wards. The patients underwent Nerve Conduction Studies at Neurophysiology Lab. The electro diagnostic criteria were used to identify patients with demyelinating or axonal types (Table 1) [15-19]. Patients were identified as falling into three major variants of GBS as AIDP, AMAN and AMSAN. Patients who didn't fall into three major variants were classified as undifferentiated. Those patients found to have other diagnoses after electro diagnosis were excluded. The NCS was performed within one week of symptom onset. Most common variant and relationship to gender and age was identified. Percentage of patients with demyelinating and axonal variants also identified. The data was analyzed using the google sheets. The permission from the institutional ethical review board was taken before starting our study.

RESULTS

A total of 180 patients were included in the study after clinical evaluation and electro diagnostic studies. Out of which 112 (62.22%) were male and 68 (37.78%) were female. The age of patients range from 6 months to 90 yrs. Mean age was 26.9±20.2. There was no significant age difference between males and females when mean age was considered.

A total of 110 (61.11%) patients had axonal type of GBS including the pure motor and mixed variety. The patients with demyelinating variety were 56 (31.11%). 14 (7.78 %) patients were of undifferentiated type.

Table No.1: Electrodiagnostic Criteria for Guillain-Barré Syndrome

	Ho et al (1995)	**Hadden et al (1998)**
Criteria for AIDP	Must have one of the following in two nerves	Must have one of the following in two nerves
CV	<90% LLN (<85%, if distal amp <50% LLN)	<90% LLN (<85%, if distal amp <50% LLN)
DML	>110% ULN (>120%, if distal amp <LLN)	>110% ULN (>120%, if distal amp <LLN)
TD	Unequivocal	Not Considered
CB	Not Considered	Proximal-to-distal amp ratio <0.5 and distal amp >20% LLN
F-wave Latency	>120% ULN	>120% ULN
Criteria for AMAN	No evidence of demyelination in the above nerves Distal amp <80% in two nerves.	None of the above except in one nerve if distal amp <10% of LLN Distal amp <80% in two nerves.

AIDP, acute inflammatory demyelinating polyneuropathy; CV, conduction velocity; LLN, lower limit of normal; DML, distal motor latency; ULN, upper limit of normal; TD, temporal dispersion; CB, conduction block; AMAN, acute motor axonal neuropathy.

Table No.2: Electrophysiologic Variants of GBS

	Total	**Males**	**Females**
AIDP	56(31.11%)	33(58.93%)	23(41.07%)
AMAN	59(32.78%)	30(50.85%)	29(49.15%)
AMSAN	51(28.33%)	37(72.55%)	14(27.45%)
Undifferen-tiated	14(7.78%)	11(78.57%)	03(21.43%)
Total	180 (100%)	112(62.22%)	68(37.78%)

Of the total patients 56 (31.11%) patients had AIDP, 59 (32.78%) had AMAN and 51 (28.33%) were of AMSAN variety. Males were affected significantly more often than females. The most common type found was AMAN (32.78%). Of which 30 (50.85%) were males and 29 (49.15%) were females. The second common type was AIDP (31.11%) of which 33 (58.93%) were males and 23 (41.07%) were females.

The third type was AMSAN (28.33%) with 37 (72.55%) males and 14 (27.45%) females. (Table 2).

DISCUSSION

Guillain-Barre syndrome (GBS) is a disorder of peripheral nerves of immune etiology which usually presents sporadically. The prevalence and incidence of different variants may vary in different regions. Although GBS is mainly diagnosed clinically, electro diagnostic studies are the basis for classification into different variants. Some studies in Pakistan and other Asian countries have identified a similar percentage of the three major subtypes. In our study AMAN variety was slightly more common than AIDP. Overall, if the pure and mixed axonal varieties are considered, 61.11% had axonal type of GBS compared to 31.11% of demyelinating variety. In our study, AIDP accounts for 31.11% of cases and AMAN accounts for 32.78% of cases. In 2006 Zaheer M et al in Pakistan in their study found 36% AIDP cases and 12% Axonal variety[14]. Another study in Pakistan showed a relatively similar pattern of GBS with demyelinating type in 46%, axonal in 31% and unclassifiable in the rest of their cases.[15] Yadegari S et al in 2014 in Iran showed that the most common type of GBS was AIDP (63%) followed by AMAN (23%) and AMSAN (14%)[2]. In 2019 in Northern China Tian J et al showed that AMAN was the most common type in 55.8% and AIDP was 21.2%[1]. A study in North India done on children with GBS found AMAN in 69.4% and AIDP in 25% children[16]. A study of Chinese patients with GBS revealed AIDP in 32% and AMAN in 55%. The higher prevalence of AMAN has been reported from China and has been attributed to Campylobacter jejuni[17].

In our region and most Middle East and Asian countries, the axonal variants of GBS seem more prevalent than North America and Europe which include only 5% of GBS cases[18]. Also the axonal variants are less common than Japan which have reported AMAN in 45-48% of their GBS cases[19].

The slight difference in the incidence of different variants is identified in different studies even in the same geographic region. This may be attributed to the type of diagnostic criteria used for classification of different variants. The criteria we used is the common criteria to classify axonal and demyelinating varieties. In many studies males are more commonly affected than females which is also shown in our study.

Our study has certain limitations. There were patients who present with rapidly worsening severe disease and are directly shifted to ICU for ventilatory support. These patients could not undergo NCS studies in the initial week. Our study does not tell us about prognosis of different variants including need of ventilatory support and time of hospital stay. The accuracy of incidence of different variants of GBS can be improved by using different electro diagnostic criteria and comparing their results. By developing a universal criteria, the true incidence of different variants can be identified in different geographic regions. This will help in better prognostication and planning for more aggressive treatment of vulnerable patients.

CONCLUSION

In our study the AMAN variant of GBS is slightly more common than the AIDP variant. The slight difference is also attributed to different criteria used to classify variants. Early identification of variants is helpful in planning treatment and prognostication of patients with severe disease.

Author's Contribution:

Concept & Design of Study:	Muhammad Wazir Ali Khan
Drafting:	Asad Hussain
Data Analysis:	Hafiz Muhammad Zeeshan
Revisiting Critically:	Muhammad Wazir Ali Khan, Asad Hussain
Final Approval of version:	Muhammad Wazir Ali Khan

Conflict of Interest: The study has no conflict of interest to declare by any author.

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