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Original Article

Electrophysiological variations of Guillain-Barre Syndrome in Bangladesh- Hospital Based Study

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Abstract:

Introduction: *Guillain-Barre syndrome (GBS) is an immune mediated disorder of peripheral nerves which usually presents by rapidly evolving ascending weakness & mild sensory loss and hypo- or areflexia. Electrodiagnostic study (EDS) is the basis for classification of different subtypes of the disease. EDS also has a crucial role in diagnosis, ruling out of some differential diagnosis like myopathic and motor neuron disorders and confirming the neuropathic nature of GBS. The benefit of immunotherapy is greatest when introduced early. In addition, electrophysiological characteristic can predict the prognosis of patients with GBS. This study was conducted to determine the predominant subtype and electrophysiological pattern of GBS in the context of Bangladesh.*

Objectives: *Objective of this study was to compare the electrophysiological variations among different electrophysiological subtypes of GBS.*

Methods: *It is an observational cross sectional study conducted in Department of Medicine and Neurology, Sir Salimullah Medical College & Mitford Hospital and National Institute of Neuroscience (NINS), Dhaka, over a period of one year & four months. Total 30 patients were selected by purposive sampling technique. Demographic data were collected from the patients and recorded in structured case report form. Clinical examination and relevant investigations were done meticulously. Collected questionnaire were checked to identify any error in data. Data was analyzed with SPSS version 21 software.*

Result: *In this study, maximum numbers of patients 53% were between 21-30 years of age group with mean value 27.47±8.1 years. Male to female ratio was 1.7:1. Frequency of Guillain-Barre syndrome is predominance at middle age group. Commonest presentation was limb weakness (parapledgia or quadripledgia) in 60% patients, paresthesias & numbness (40%), pain (100%) and deviation of mouth (63%) of GBS patients. Cerebrospinal fluid shows a mild pleocytosis (5 to 50 cells/µl) in majority of cases was found in 76% of patients. Whereas elevated CSF Protein (>45 mg/dl) was seen in all GBS patient. Increased distal motor latency (DML) was found in 93% patients, whereas 7% patients had normal DML. In the case of the lower limbs, increased distal motor latency was predominant. Decreased amplitude of sensory nerve action potential (SNAP) was seen in 83% patients while 16% patients had normal. Slowing of motor conduction velocities, decreased amplitude as well as increase in distal motor latencies were observed, being more pronounced in the lower limbs. F-wave was completely absent in 20.0% patients while 20% patients showed decreased conduction velocity with prolonged latency. Sensory nerve action potential revealed that decreased sensory conduction velocity (SCV) was seen in 26%, absent SCV in 10%*

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and normal SCV in 63% patients. Present study demonstrated that Acute inflammatory demyelinating polyneuropathy (AIDP) was the commonest type of Guillain-Barré syndrome, present in 56% of patient. Around 30% of the patients belonged to acute motor axonal polyneuropathy (AMAN) and 13% were acute motor sensory axonal polyneuropathy (AMSAN).

Conclusion: In this study AIDP was the most frequent subtype. The characteristic 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.*

Key words: GBS, electrophysiology, AIDP, AMAN

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Introduction:

Guillain-Barré Syndrome (GBS) poses a formidable challenge within the realm of neurological disorders, emerging as a leading cause of acute flaccid paralysis on a global scale, particularly in the post-poliomyelitis vaccine era. This acute, often severe polyradiculoneuropathy has autoimmune origins, with its roots in acute inflammatory demyelinating polyneuropathy (AIDP) recognized over a century ago. Initially characterized by an immune assault on peripheral nerve myelin and consequential axon loss, the landscape has expanded to include axonal motor and sensorimotor variants, influenced by molecular mimicry targeting motor axons[1].

Clinical diagnosis heavily relies on meticulous history-taking and examination, complemented by supportive evidence from cerebrospinal fluid analysis and electrodiagnostic testing[2]. The syndrome's initial symptoms commonly manifest as acroparesthesia with minimal objective sensory loss and severe radicular back pain. Differential diagnosis considerations involve a wide range of neurologic assessments to localize the disease to peripheral nerves, distinguishing it from conditions affecting the brain, spinal cord, cauda equina, neuromuscular junction, or muscles[3].

Geographically, GBS incidence and subtype distribution vary, emphasizing the need for a nuanced approach to diagnosis and classification. Approximately 60% of GBS cases globally are preceded by viral or bacterial infections, highlighting a potential link between infections and the immune system's aberrant response[4].

GBS significantly impacts the peripheral nervous system, leading to muscle weakness, sensory loss, cranial nerve weakness, diaphragmatic weakness, and autonomic disturbances[5]. The immune system's misguided assault on nerve roots and peripheral nerves results in defects in nerve impulse propagation, culminating in flaccid paralysis. Recent insights have revealed GBS's heterogeneity with distinct pathological entities, including predominantly axonal patterns like acute motor-sensory axonal neuropathy (AMSAN) and acute motor axonal neuropathy (AMAN)[6]. Electromyography (EMG) and nerve conduction studies (NCS) aid in diagnosis, showcasing abnormalities consistent with demyelination.

In Bangladesh, a tertiary center hospital study sought to unravel the predominant GBS subtypes and compare nerve conduction studies between axonal and demyelinating forms. The findings from this research contribute to the global understanding of GBS, shedding light on the nuances of its presentation and the implications for electrodiagnostic testing.

In this comprehensive exploration of GBS, we embark on a journey through its clinical, pathological, and diagnostic intricacies, aiming to unravel the mysteries that surround this autoimmune polyradiculoneuropathy.

Material and Methods

This observational cross-sectional study, conducted at Sir Salimullah Medical College & Mitford Hospital, Dhaka, and the National Institute of Neuroscience (NINS), Dhaka, spanned 1 year and 4 months (January 1, 2017, to April 30, 2018). Targeting 30 consecutively diagnosed Guillain-Barré Syndrome (GBS) patients, the sample size was determined using $N = Z^2 \frac{2pq}{d^2}$ formula. Purposive sampling enrolled both genders meeting GBS diagnostic criteria, with inclusion involving progressive motor weakness in more than one limb and areflexia, and exclusion excluding sensory involvement, lack of electrophysiological data, unwillingness to consent, metabolic abnormalities, and age below 12.

Operational definitions were set for GBS subtypes (AIDP, AMAN, AMSAN), diagnosed using CSF analysis and NCS based on Dutch Guillain Barre study group criteria. Data collection involved structured forms covering demographics, clinical presentation, comorbidities, and investigations, including NCS. SPSS analyzed the processed data ($p<0.05$ significance level). Quality assurance included a manual, pretesting, ethical approval, and confidentiality assurance. The study protocol was ethically approved, with written consent obtained from patients or relatives, ensuring information confidentiality throughout.

Result:

Table-1: *Distribution of respondents according to age (n=30)*

Age (years)	Frequency Male (n, %)	Female (n, %)	Total (%)	Mean ± SD
<20	4(21.0)	0	4(13.3%)	
21-30	8(42.1)	7(63.6)	16(53.3%)	
31-40	5(26.3)	3(27.2)	7(23.3%)	27.47±8.1
41-50	2(10.5)	1(9.0)	3(10.0%)	
Total	19	11	30(100.0)	

In this study, maximum numbers of patients 16(53%) were between 21-30 years of age group, next 7(23.3%) were between the age group of 31-40 years, with mean value 27.47±8.1 years. Frequency of Guillain-Barre syndrome is predominance at middle age group. Out of 30 cases 63% patients were male and 37% were male. Male to female ratio was 1.7:1. In case of male and female 21-30 years was having highest incidence and female patients were comparatively more in older age. Study also showed large numbers of respondents came from urban area (60.0%), followed by rural area (30.0%) and sub-urban/slum area was 10.0%. Maximum of respondents were daily worker (40%) followed by housewife (23%). A considerable portion of the respondents (13%) were service holder. We observed that most of the female patients were housewives.

In this study we found that majority of patients 25(83.3%) had history of illness for <4 weeks. Present study shows that limb weakness (parapledgia or quadripledgia), paresthesias & numbness, pain and deviation of mouth were the commonest presentation in GBS patients (60.0%, 40.0%, 100.0% & 63.3% of patients respectively). Other clinical manifestations were respiratory distress in (46.6%) of cases, dysphagia (60%), visual disturbance (43%) of cases. And among the clinical sign majority of patients found tachycardia (46.6%) and hypertension (50%), followed by Shortness of breath (46.6%), Facial flushing (30%) and planter extensor (33.%).

Table-2: *Distribution of respondents according to clinical manifestation*

Presentation	Frequency	Percentage (%)
Parapledgia	18	60.0
Quadripledgia	12	40.0
Paresthesias, numbness	30	100.0
Deviation of mouth	19	63.3
Pain	23	76.6
Respiratory distress	14	46.6
Dysarthria	9	30.0
Dysphagia	18	60.0
Convulsion	5	16.6
Diplopias	8	26.6
Visual disturbance	16	53.3
Ophthalmoplegia	8	26.6
Sphincter problem	18	60

N.B. Multiple respondents

Table-3: *Distribution of respondents according to physical sign*

Sign	Number of patients	Percentage (%)
Tachycardia	14	46.6
Bradycardia	11	36.6
Shortness of breath	14	46.6
Facial flushing	9	30
Hypertension	15	50
Hypotension	8	26.6
Papilloedema	7	23.3
Planter extensor	10	33.3

N.B. Multiple respondents

Present study demonstrated that, Cerebrospinal fluid shows a mild pleocytosis (5 to 50 cells/µl) in majority of cases was found in 23(76.6%) of patients. Whereas elevated CSF Protein (>45 mg/dl) was seen in all GBS patient.

Increased distal motor latency (DML) was seen in 28 (93%) patients, 2(7%) patients had normal DML. On the case of the lower limbs, increased distal motor latency was

Table-4: *Distribution of respondents according to electrophysiological findings (n=30)*

Nerve conduction test (NCV)	Variables	Frequency	Percentage (%)
F- waves Abnormal	Absent	6	20.0
	Prolonged latency	6	20.0
	Normal	0	0
Sensory conduction velocity (SCV)	Absent	3	10.0
	Decreased	8	26.6
	Normal	19	63.3
Amplitude of sensory nerve action potential (SNAP)	Decrease	25	83.3
	Normal	5	16.6
Distal motor latency (DML)	Increase	28	93.3
	Normal	2	6.6
Conduction block		8	26.6
Compound muscle action potential (CMAP)	Decrease	26	86.6
	Normal	4	13.3

N.B. Multiple respondents

predominant. Decreased amplitude of sensory nerve action potential (SNAP) was seen in 25(83.3%) patients while 5(16.6%) patients had normal. Slowing of motor conduction velocities, decreased amplitude as well as increase in distal motor latencies were observed, being more pronounced in the lower limbs. F-wave was completely absent in 6(20.0%) patients while 6 patients (20.0%) showed decreased conduction velocity with prolonged latency. Sensory nerve action potential revealed that decreased sensory conduction velocity (SCV) was seen in 8(26.6%), absent SCV in 3(10.0%) and normal SCV in 19(63.3%) patients.

Table-5: *Distribution of respondents according to nerve conduction study (n=30)*

Nerve conduction studies	Frequency	Percentage
Demyelinating	17	56.6
Axonal	12	40.0
Equivocal	1	3.33

EMG studies were carried out in 30 patients of the GBS, of which 17(56.6%) showed demyelinating type of polyneuropathy with reduced voluntary motor unit recruitment while the remaining 12(40.0%) patients showed axonal type and 1(3.3%) equivocal polyneuropathy.

Present study demonstrated that AIDP was the commonest type of Guillain-Barré syndrome, present in 56.6% of patient. Around 30.0% of the patients belonged to AMAN and 13.4% were AMSAN.

The most frequent inexcitable motor nerve was peroneal tibial nerve. In AIDP patients, prominent CMAP amplitude

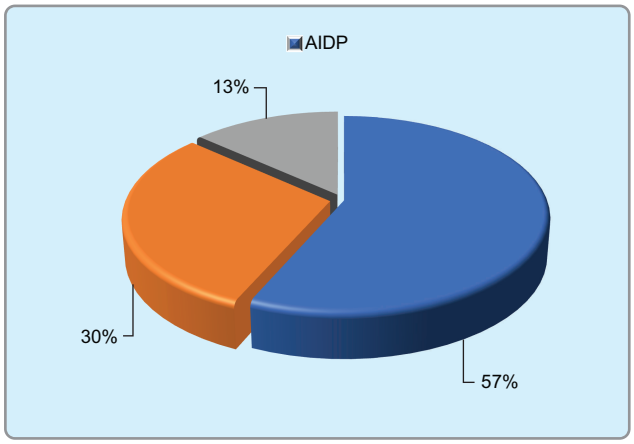

Figure-1: *Distribution of respondents according to electrophysiological types (n=30)*

AIDP: Acute inflammatory demyelinating polyneuropathy; AMAN: Acute motor axonal polyneuropathy, AMSAN: Acute motor sensory axonal polyneuropathy

reduction was observed in tibial nerves, while in median nerve mild reduction occurred. CMAP amplitude (mV) was 2.8 & 3.3 in AMAN & AMSAN respectively. Motor DL preserved in AIDP, but in AMAN was 3.5 and 3.8 ASMAN.

In this study most frequent inexcitable sensory nerve was sural nerve. Unexcitable nerves were more common among examined sensory nerves, especially those evaluated at late stage. The reason is probably related to the time that takes Wallerian degeneration occurs, which is longer for sensory than motor nerves and subsequently results in SNAP amplitude reduction to its nadir later than CMAP amplitude. In this study sensory NCS, prominent discrepancy between upper and lower limbs was seen in electrophysiological findings: SNAP (Amplitude of sensory nerve action potential) was comparably reduced in both upper and lower limbs in AIDP, but sural SNAP amplitude showed slightly preservation than median SNAPs. The median of SNAP amplitude remained normal in AMAN, but SNAP amplitudes revealed more reduction in lower limb.

Table shows electrophysiological features of GBS patients. Unobtainable H-reflex was the late response with the greatest frequency of abnormality. Abnormal late responses were obviously common in demyelinating rather than axonal GBS. F-response was abnormal in most of cases. In AIDP, the most frequent abnormality of F-response was prolonged F-wave latency (35.2%) and in AMAN none case detected. Absent F-wave was detected in 20.0% of patients (4 patients with AIDP and 2 with AMAN). The frequency of sural sparing was 29.4% in AIDP.

Table- 6: *Distribution of respondents according to results of motor nerve conduction study (n=30)*

Nerve	Normal/ Control median (range)	AIDP (n=17) median (range)	AMAN (n=9) median (range)	AMSAN (n=4) median (range)
Median				
CMAP amplitude (mV)	≥4.0	3(0-8)	2.8(0-11)	3.3(1-11)
Motor DL (ms)	≤4.4	6.2(3-12)	3.5(3-5)	3.8(3-6)
NCV (m/s)	≥49	45	53	48
Tibial				
CMAP amplitude (mV)	≥4	1.3(0-5.0)	1.5(0-8)	1.5(0-5)
Motor DL (ms)	≤5.8	6.7(0-14.3)	7.2(5-8)	5(5.0-5.1)
NCV (m/s)	≥41	28(0-71)	42(0-46)	39(0-46)

Data are presented in median (range). AIDP: Acute inflammatory demyelinating polyneuropathy; AMAN: Acute motor axonal polyneuropathy, AMSAN: Acute motor sensory axonal polyneuropathy; NCV: Nerve conduction velocity; DL: Distal latency, CMAP: Compound muscle action potential.

Table-7: *Distribution of respondents according to results of sensory nerve conduction study (n=30)*

Nerve	Normal/ Control median (range)	AIDP (n=17) median (range)	AMAN (n=9) median (range)	AMSAN (n=4) median (range)
Median				
Amplitude (iV)	≥20	6(0-84)	45(0-72)	10.3(0-42)
NCV (ms)	≥50	48.7	56.2	53.1
Sural				
Amplitude (iV)	≥6	3.7(0-18)	12.5(4-28)	0
NCV (ms)	≥40	38	45	0

Data are presented in median (range). AIDP: Acute inflammatory demyelinating polyneuropathy, AMAN: Acute motor axonal polyneuropathy; AMSAN: Acute motor sensory axonal polyneuropathy; NCV: Nerve conduction velocity.

Table-VIII: *Distribution of respondents according to electrodiagnostic variation in different types of Guillain-Barre syndrome (n=30)*

Electrophysiological feature	AIDP (n=17)	AMAN (n=9)	AMSAN (n=4)
Conduction block	7(41.1%)	1(11.1%)	0
Temporal dispersion	5(29.4%)	0	0
Absent F-wave	4(23.5%)	2(22.9%)	0
Prolonged F-wave	6(35.2%)	0	0
Unobtainable H reflex	13(76.4%)	2(22.9%)	0
Sural sparing	5(29.4%)	0	0

Discussion:

This study delves into the epidemiological, clinical, and diagnostic aspects of Guillain-Barré Syndrome (GBS), uncovering noteworthy patterns. The investigation revealed a peak incidence of GBS in the 21-30 age group, with a male-to-female ratio of 1.7:1, mirroring global trends[1]. The middle age group exhibited a GBS prevalence, consistent with findings in other studies[5], and geographical variations were absent, aligning with the understanding that GBS affects individuals of all ages, with a higher occurrence in adults and males.

Clinical presentations predominantly featured limb weakness, paresthesias, pain, respiratory distress, dysphagia, and visual disturbances. Limb weakness, progressing symmetrically in an ascending or descending pattern, was a hallmark, with a majority reporting illness lasting less than four weeks, emphasizing the acute nature of GBS. This aligns with established literature underlining the significance of early diagnosis in rapidly progressing paralysis cases[7].

Crucially, cerebrospinal fluid (CSF) analysis played a pivotal role in supporting GBS diagnosis, revealing mild pleocytosis and elevated protein levels in the majority. 'Cyto-albuminologic dissociation' in CSF, characteristic of GBS, manifested with heightened sensitivity after a week of weakness, underscoring the diagnostic utility of CSF analysis, particularly in uncertain cases.

Electrophysiological findings highlighted increased distal motor latency and decreased amplitude of sensory nerve action potential, more pronounced in lower limbs. This corresponds with established electrodiagnostic criteria, emphasizing the role of nerve conduction studies in confirming GBS diagnosis.

The study classified GBS into subtypes, with Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP) being the most common, followed by Acute Motor Axonal Neuropathy (AMAN) and Acute Motor Sensory Axonal Neuropathy (AMSAN). This variability in clinical and electrophysiological phenotypes aligns with existing literature, showcasing the diverse spectrum of GBS manifestations[8].

While the study provides comprehensive insights into adult GBS features, limitations such as a small sample size and a single-center approach are acknowledged. Despite these, the findings underscore the importance of early clinical recognition, electrophysiological confirmation, and CSF analysis in GBS diagnosis. This study contributes valuable knowledge, yet larger cohorts and multi-center studies are recommended for a more comprehensive understanding of GBS manifestations, refining diagnostic and management strategies.

Conclusions:

The diagnosis of GBS and defining its subtypes should not be made based on a single finding and clinical features, CSF profile and electrodiagnostic evaluation should be considered together.

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