

Electrophysiology in the Guillain-Barré Syndrome: Study of 30 Cases

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Summary:

Thirty consecutive patients diagnosed clinically as Guillain Barré Syndrome (GBS) were enrolled in this study to see the electrophysiological patterns of GBS in Bangladeshi community. Among 30 patients, 25 were male (M: F = 5:1) and 47% patients were between 16 and 25 years of age. An antecedent event was present in 67% of patients. An elevated protein was present in 90% of cases and a cell

count of up to five was present in 94% of patients. Acute inflammatory demyelinating polyradiculopathy (AIDP) was commonest (33.35%) followed by acute motor axonal neuropathy (AMAN) which constitute 26% of patients in electrophysiological study of the enrolled patients. Acute motor sensory axonal neuropathy constitutes 14% of cases in this series.

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Introduction:

The term Guillain-Barre syndrome (GBS) defines a recognizable clinical entity that is characterized by rapidly evolving symmetrical limb weakness, loss of tendon reflexes, absent or mild sensory signs, and variable autonomic dysfunction¹. Guillain-Barre syndrome is now the commonest cause of acute neuromuscular paralysis worldwide affecting patients of all ages and both sexes².

The earliest descriptions of what we know today, as Guillain-Barre syndrome are probably that of Wardrop and Ollivier, in 1834. In 1916 Guillain, Barre, and Strohl described two French soldiers with motor weakness, areflexia and "albuminocytological dissociation" in the cerebrospinal fluid³. Subsequently several cases with similar manifestations were reported and this clinical entity was named after Guillain and Barre. The Guillain-Barre syndrome, as recognized clinically, was frequently been equated with acute inflammatory demyelinating polyradiculopathy (AIDP), the pathological entity most extensively studied and most frequently reported in older studies⁴⁻⁵ and in contemporary reports⁶⁻⁷. However, several lines of evidence suggest that the pathological basis for the Guillain-Barre syndrome, as clinically diagnosed, can be broader, as recently reviewed⁸⁻⁹. Some suggests that axonal degeneration can be the dominant process¹⁰⁻¹¹. In the United States, Europe and Australia, the predominant form is AIDP. The concept of axonal

Guillain-Barre syndrome was first raised by Feasby et al, who found early axonal degeneration of motor and sensory fibers in five patients with clinically defined Guillain-Barre syndrome. This pattern of Guillain-Barre syndrome, termed acute motor sensory axonal neuropathy (AMSAN), is usually associated with a worse prognosis than demyelinating Guillain-Barre syndrome. Studies in northern China and India¹² have identified another form of axonal Guillain-Barre syndrome, termed acute motor axonal neuropathy (AMAN)¹³. It is suggested that AMAN is associated with pure motor axonal involvement, anti-ganglioside GM1 antibodies, or preceding *Campylobacter jejuni* infection¹⁴⁻¹⁶. Chinese patients with AMAN are reported to often develop hyperreflexia during the early phase of recovery¹⁷. Preserved or exaggerated tendon reflexes do not usually occur in patients with AIDP or in patients with other peripheral neuropathies.

F wave represent method of looking at the proximal segments of motor nerves and as such can be very important in certain circumstances (e.g. GBS). The absence of F wave has been interpreted as demyelinating conduction block in the proximal nerve segments when distal compound muscle action potentials are preserved. In Guillain-Barre syndrome, peripheral nerve conduction studies are normal in 10% to 20% of patients. However, electrophysiological evidence of nerve dysfunction may be present in the proximal portion of the peripheral nerves and thus absence or slowing of F wave may be an isolated conduction abnormality, especially in the early stage of illness.

Materials and method:*Place of study:*

This study was carried out in the department of neurology, Bangabandhu Sheikh Mujib Medical University (BSMMU) Hospital during the period of July 2000 to March 2002.

Type of study:

This was a prospective clinical and electrophysiological study.

Inclusion criteria:

Patients who met the diagnostic criteria for Guillain-Barre syndrome (Asbury and Cornbath, 1990) as describe below were included in this study.

- I. Features required for diagnosis:
 - A. Progressive motor weakness of more than one limb.
 - B. Areflexia.
- II. Features strongly supportive of the diagnosis:
 - A. Clinical:
 1. Progression within four weeks
 2. Relative symmetry
 3. Mild sensory symptoms or signs
 4. Cranial nerve involvement
 5. Recovery within four weeks of progression stopping
 6. Autonomic dysfunction

Subject: Thirty consecutive patients (25 male and five female) were included in this study. The youngest patient in this series was a 12 years old boy and the eldest case was 67 years old male.

Method: After admission into the Neurology department of BSMMU Hospital, a thorough history including the history of any antecedent events was taken from the patient or patient's attendant. In every case a careful neurological examination was done including the autonomic nervous system. Patients were followed up for respiratory function, blood pressure, pulse, and muscle power. Each of the patients, who were diagnosed clinically as GBS, was investigated for confirming diagnosis by doing cerebrospinal fluid (CSF) study and electrophysiological study. CSF study

was done after tenth day of illness and was sent for biochemical, bacteriological and cytological examination. Electrophysiological study was done irrespective of duration of illness. All measurements were done with surface electrodes and measurements were recorded in a form used by Neurology department in BSMMU. Nerves were stimulated using 1ms electrical pulses at a repetition rate of one per second with intensity sufficient to elicit maximum amplitude of compound muscle action potential (CMAP) and sensory nerve action potential (SNAP). In addition to distal latency, amplitude and nerve conduction velocity nerves were tested for F wave. Besides these, some other investigations were done for example, serum electrolytes, blood sugar etc. to exclude any possible secondary causes of muscle weakness, Data were collected by using a questionnaire and analyzed by appropriate statistical method.

Operational definition at electrophysiology:

Acute Inflammatory Demyelinating Polyneuropathy (AIDP):

Reduced conduction velocity;
Conduction block or temporal dispersion;
Prolonged terminal latency; and
Absent F wave or prolonged F wave latency.

Acute Motor Axonal Neuropathy (AMAN):

Absent or reduced compound muscle action potential (CMAP);
Normal motor terminal latency and conduction velocity; and
Normal sensory nerve action potential (SNAP).

Acute Motor Sensory Axonal Neuropathy (AMSAN):

Absent or reduced SNAP amplitude;
Absent or reduced CMAP amplitude; and
Normal motor terminal latency and nerve conduction velocity.

Results:

The study population consisted of thirty (30) patients, twenty-five were male and five female. The age distribution of patients is shown in Table-I. The youngest case in this series was a 12 year old boy and the eldest was a 67 year old male. However, most

(46.67%) of the patients were in between 16 and 25 years of age. A history of antecedent event was present in 19 (66.33%) cases while 11 (33.66%) patients failed to give any preceding history of an antecedent event. Fever alone or in combination with loose motion was the commonest antecedent event (36.66%) followed by loose motion (23.33%). No associated condition like trauma, surgery, lymphoma, vaccination history was present in this study group.

Table-I

Showing the distribution of patients according to age groups (N=30)

Age group in years	No of patients
0-15	03 (10%)
16-25	14 (46.65%)
26-35	03 (10%)
36-45	06 (20%)
46-60	03 (10%)
>60	01 (3.33%)

Seventh cranial nerve was the commonest nerve (36.66%) involved in this series and it was bilateral in two third cases. Sensory involvement was present in five (16.60%) patients in the form of impairment of pain and touch sensation. Autonomic involvement like tachycardia, bradycardia, hypertension, hypotension or constipation was present in 27% cases.

Cerebrospinal fluid protein was elevated in 90% patients. In 28 (93.3%) cases cerebrospinal fluid total

cell count was up to 5 cells/mm³ and in only two (6.7%) patients total cell count was up to 15 cells/mm³ (Table-II).

Table-II

Showing results of CSF study in Guillain-Barré syndrome patients

Parameter	Elevated	Normal
Protein	27 (90%)	3 (10%)
Cell count	2 (6.70%)	28 (93.30%)

The commonest pattern (33.33%) in electrophysiology was AIDP followed by AMAN and combination of AMAN and AIDP patterns, both constitute about 26.66% of cases. Acute motor sensory axonal neuropathy constitute about 14% of patients (Table-III). In Table-IV and Table - V details of nerve conduction study and findings of CSF examination of selected patients are presented.

Table-III

Showing electro physiologically defined groups of Guillain-Barré syndrome

Electro physiologically defined group	Number of patients
AMAN	04 (13.33%)
AMSAN	08 (26.66%)
AIDP	10 (33.33%)
AMAN and AIDP	08 (26.66%)

Table-IV

Showing the figures of nerve conduction study (NCS) at median nerve of few Guillain-Barré syndrome cases

Case No	NCS Summary	Motor Study			Sensory Study
		NCV	Amplitude	F wave	Amplitude
1	Axonal (motor) & demyelinating	50.23 msec	5.5 mv	Absent	17.1 mv
2	Axonal motor sensory	52.10 msec	4.9 mv	26.3	8.50 mv
3	Demyelinating	32.80 msec	12mv	Absent	18.20 mv
5	Axonal motor	52.10 msec	4.7 mv	26.7	20.21 mv
13	Demyelinating	37.23 msec	11.4 mv	27.1	17.2 mv
22	Axonal (motor) & demyelinating	51.12 msec	4.7 mv	41.24	17.4 mv
24	Axonal motor	52.15 msec	4.6 mv	26.15	21.5 mv
30	Demyelinating	39.12 msec	12.10 mv	Absent	14.20 mv

Table-V

Showing the summary of nerve conduction study at median nerve and figures of CSF study of Guillain-Barré syndrome cases

Case No.	NCS Summary	CSF Study		Sex	Age
		Protein (mg/dl)	Days from onset		
1	Axonal (motor) and demyelinating	100	12	M	32
2	Axonal motor sensory	180	13	M	20
3	Demyelinating	200	10	M	56
5	Axonal motor	75	12	M	60
9	Demyelinating	28	15	M	33
22	Axonal (motor) and demyelinating	145	11	M	8
23	Demyelinating	136	12	F	25
30	Axonal motor	32	14	M	36

F wave was absent or F wave latency was prolonged in four (50%) patients among eight cases in whom it was done within seven days of onset of illness. F-wave parameters were abnormal in eight (36.36%) cases among patients who presented after seven days of onset of the disease. Compound muscle action

potential (CMAP) was abnormal in three cases (37.50%) out of eight patients who presented within seven days of illness, and in 17 (77.27%) patients out of 22 CMAP were of low amplitude when it was done after seven days (Table-VI).

Table-VI

Evolution of nerve conductive study changes in Guillain-Barré syndrome cases

Changes	Day 1 to 7 (n=8)	Day 7 to 21 (n=22)	Total number of patients (n=30)
F waves:			
Abnormal	04 (50%)	08 (36.36%)	12 (40%)
Absent	03 (75%)	06 (75%)	09 (75%)
Prolonged	01 (25%)	02 (25%)	03 (25%)
Normal	04 (50%)	12 (63.64%)	16 (60%)
SNAP response:			
Abnormal	01 (12.5%)	04 (18.18%)	05 (16.67%)
CMAP response:			
Low amplitude			
>2 Nerves	03 (37.5%)	17 (77.27%)	20 (66.67%)

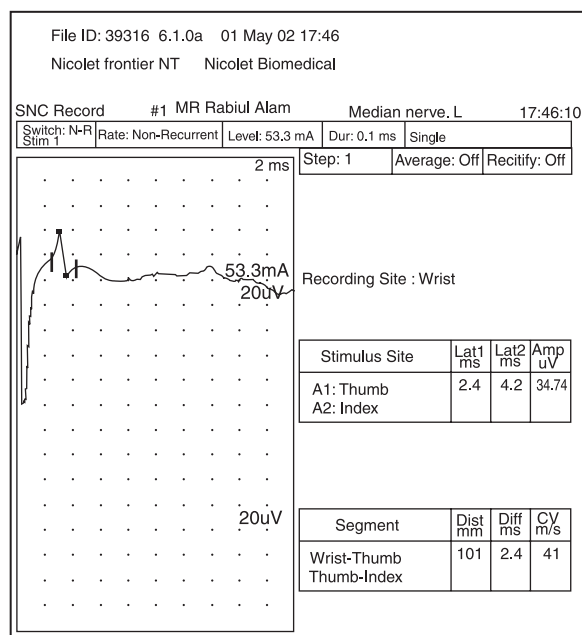


Fig.-1: Showing sensory nerve conduction in a patient of Guillain Barre syndrome.

Discussion:

The present study was undertaken to evaluate the electrophysiological changes in Guillain-Barre syndrome patients in Bangladeshi population. The standard values used at BSMMU electrophysiology laboratory were taken as control.

Male patients outnumbered female patients, as this study showed males suffered as much as five times (M: F=5:1). Though some studies showed males appeared to be affected more commonly¹⁸⁻¹⁹, the outstanding male predominant involvement in this study was quite remarkable. This may be a chance finding or may be explained on the facts that female patients fail to reach to a tertiary center, like this, for treatment because of customs and religious background or other social factors.

The age distribution curve showed 47% of patients were between 16 and 25 years of age. The next common age of presentation was between 36 and 45 years (20%). This was corroborated with other studies where the investigators showed that the young adults between 15 and 25 years are peak age group for suffering from Guillain-Barre syndrome^{18, 20}.

The presence of antecedent events was present in two third of patients (67%) in the preceding weeks (one to

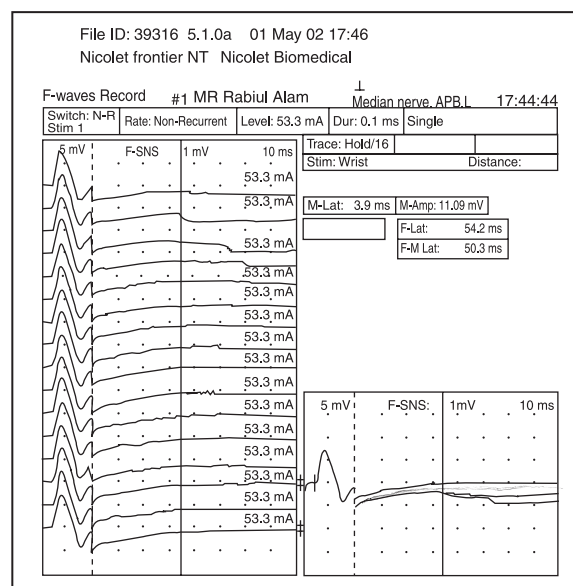


Fig.-2: Figure-2 : Showing F-waves recording of a patient of Guillain-Barré syndrome.

four weeks) and this finding was well in association with other studies²¹⁻²².

Cranial nerve involvement was present in 46.66% cases and seventh cranial nerve was the commonest (78.50%) and it was bilateral in two third cases. This finding is consistent with two other studies,²³⁻²⁴ where cranial nerve involvement was present in 55% and 53% of cases respectively.

Features of autonomic nerve involvement such as sinus tachycardia and bradycardia, hypertension and postural hypotension, constipation, anhidrosis or excessive sweating were present in 27% of patients. This finding is quite low when compared with the study carried out by Singh et al where they showed two third of their cases had features of autonomic nerve involvement²⁵. However, these features should be considered carefully during patient management to avoid catastrophe.

Cerebrospinal Fluid study revealed presence of elevated protein in 90% of cases, which were more or less well correlated²². The cell count in cerebrospinal fluid was up to 5 cells/mm³ in 94% of patients and in only 6% of cases it was up to 15 cells/mm³. This study restated the old saying "cyto-albumino dissociation" is a characteristic feature of GBS.

F wave was absent or F wave latency was prolonged in four (50%) patients among eight cases in whom it was done within seven days of onset of illness. F-wave parameters were abnormal in eight (36.36%) cases among patients who presented after seven days of onset of the disease. Thus this study showed that F-wave abnormalities are more useful in assessing Guillain-Barre syndrome patients in early days.

Acute inflammatory demyelinating polyradiculopathy is the most prevalent form of GBS in western countries and accounts for 85-90% of cases²⁶. AIDP was found in 85% of cases and acute axonal forms of GBS in 15% of cases in a series reported by Hahn²⁷. In this series, AIDP was the commonest electrophysiological variant of GBS (33.35%). This is quite low in comparison to the western studies as mentioned³⁻⁶. This study revealed that in 14% cases there were presence of both motor and sensory axonal damage and this finding was well correlated with other published reports²⁸.

In this study, AMAN and a combination of AMAN and AIDP closely follows AIDP, each group constituted about 26% of cases. This was slightly higher than other reported studies especially from China²⁸⁻²⁹. That is in about, 26% cases there were presence of both demyelination and axonal changes in the same patient.

As axonal regeneration takes long time, the functional recovery after Guillain-Barré is delayed in whom the axon is the main target of damage. On the other hand remyelination occurs much quickly and thus the chance of recovery following GBS is much better in whom only myelin sheath is involved. Thus the ultimate outcome depends mainly on the pattern of involvement.

This study shows that acute inflammatory demyelinating neuropathy is still the predominant GBS variant in Bangladeshi population but axonal involvement and injury constitute a substantial percentage of GBS cases, which should be sought in clinical and electrophysiological study to foresee the ultimate outcome in GBS.

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