

Early Electro diagnostic Findings of Guillain Barre Syndrome: Case Series

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Abstract

Introduction: Guillain Barre Syndrome (GBS) is an acute immune mediated de-myelinating poly-radiculo-neuropathy. As early diagnosis favors a good outcome after treatment, this study was carried out to analyze the electro-physiological abnormalities in the first week of GBS in this region. **Case presentation:** The study was carried out for early confirmation of GBS in 65 clinically diagnosed patients reporting within a week of muscle weakness by electro-physiological tests in the years 2010-12. **Conclusion:** Slowing of motor conduction velocities, decreased amplitude and increased distal motor latencies (DML) was seen in peroneal, tibial, median and ulnar nerves in descending order of severity. Conduction block was seen in the lower limb in 10 (15.38%) patients. F-wave was completely absent in upper and lower limbs in 50 (76.92%) patients while 15 patients (23.07%) showed decreased F-wave conduction velocity in both limbs. Sensory conduction velocity of median nerve was less compared to sural nerve. Electromyography (EMG) studies showed that de-myelinating type of neuropathy was predominant (83.33%). Electro-physiological studies play an important role in the early detection; characterization & treatment of GBS because timely intervention reduces morbidity and disability. Increased DML, absent F- wave, decreased median with normal Sural SCV (sensory conduction velocity) is diagnostic of early GBS.

Key words: Guillain Barre Syndrome, Peripheral neuropathy, de-myelinating poly-radiculo-neuropathy, Electromyography

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Introduction

GBS is an auto-immune mediated de-myelinating polyradiculo-neuropathy. Males & females are equally at risk. Clinical features include progressive, symmetrical ascending muscle weakness of more than two limbs, areflexia with or without sensory, autonomic and brainstem abnormalities. Weakness is prominent in leg muscles as compared to arms; there is absence of fever at the onset of neural symptoms. Cranial nerve involvement may affect airway and facial muscles, eye movements and swallowing¹. It usually presents with numbness and tingling in the feet².

In 1949, Haymaker & Kernohan reported the histo-pathological features of 50 fatal cases of GBS. The earliest features were edema of proximal nerves followed by degeneration of myelin sheath within the 1st week of illness³

Electro-diagnosis plays an important role in early detection and characterization of inflammatory de-myelinating polyradiculopathies⁴.

Nerve conduction abnormalities become more prominent during the initial weeks of the disease even if patients clinical status is

improving^{5,6}. Early nerve conduction findings include abnormal or absent F waves with low CNAP's, an abnormal upper extremity sensory nerve action potential combined with normal sural response and multiple indirect discharges^{4,7,8}.

Case report

Ethics committee approval: There was no issue of ethical committee approval during this study as the patients were referred from department of Medicine of our Institute for electro-diagnostic evaluation.

The present study was carried out to facilitate early confirmation of clinically diagnosed cases of GBS in the 1st week of illness that were sent to the Department of Physiology for nerve conduction. The study was conducted on these 65 subjects (42 males & 23 females) between the age group of 6-70 years using RMS EMG EP Mark-II Chandigarh (Table 1). The criteria for clinical diagnosis of GBS were rapidly progressive limb weakness with or without distal limb paresthesias and reduced deep tendon reflexes.

Table 1: Clinical features in 65 patients of GBS:

Variables	Number of Patients
Males/Females	42/23
Age Range	6-70
Age<60 years	59
Weakness	65/65
Areflexia	58/65

The parameters considered were divided into four major groups –

1. Motor conduction studies included: Median, ulnar, tibial and peroneal conduction velocities, amplitude and their distal motor latencies.
2. Sensory conduction studies included: Medial and sural nerve conduction velocities.
3. F wave studies: F wave studies included F wave conduction velocity & F wave latency. (F wave is a late response resulting from anti-dromic activation of motor neurons involving conduction to and from spinal cord. F wave studies have been established as a valuable tool in clinical neuro-physiology⁹. Prominent slowing of F waves has been reported in GBS where the demyelination may affect the proximal segment of nerve and even the roots which

cannot be assessed by routine nerve conduction studies¹⁰.

4. EMG studies (Electro-myographic studies): conducted in Deltoid & Abductor digiti minimii (Upper Limb) and Tibialis anterior & Peroneus longus muscles (lower limb).

Electro-diagnostic criteria:

(According to Dutch Guillain Barre study group criteria) only one of the following abnormalities in at least two nerves should be considered:¹¹

1. Increased distal motor latency > 150% of upper limit of normal
2. Decreased conduction velocity < 70% of lower limit of normal
3. Increased F wave latency > 150 of upper limit of normal
4. Decreased compound muscle action potential amplitude > upper limit of normal.

The placement of electrodes was done on the basis of the technique described by Mishra & Kalita¹².

Results :

Distal motor latency:

- Increased distal motor latency (DML) was seen in 42 (64%) patients for both ulnar & median nerves and exclusively

for ulnar nerve in 8 (12%) patients. 15 (23.07%) patients had normal DML. On the case of the lower limbs, increased distal motor latency was seen in 50 (76.92%) patients for both tibial & peroneal nerves and exclusively for tibial nerve in 5 (7.69%) patients.

Conduction velocity & amplitude:

Decreased conduction velocity & amplitude was seen in 44 (67.6%) for both median & ulnar nerves and exclusively for ulnar nerve in 8 (12.3%) patients while 13 (20.1%) patients had normal conduction velocities. In lower limbs, decreased conduction velocity & amplitude was seen in 50 (76.92%) for both tibial & peroneal nerves and exclusively for tibial nerve in 5 (7.69%) patients. Conduction block was seen in the lower limb in 10 (15.38%) patients.

So, 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: (Table 2)

F-wave was completely absent in both upper and lower limbs in 50 (76.92%) patients while 15 patients (23.07%) showed decreased conduction velocity with

prolonged latency and decreased amplitude in both upper and lower limbs.

Sensory Nerve action potential: (Table 2)

Decreased sensory conduction velocity (SCV) was seen in 40 (61.29%), absent SCV in 20 (30.76%) and normal SCV in 5 (7.69%) patients in the upper limb. On the other hand, normal Sural nerve SCV [SSCV] was seen in 55 (84.61%), decreased SSCV in 5 (7.69%) and absent SSCV in 5 (7.69%) patients in the lower limb.

So, sensory conduction velocity of median nerve was slightly less as compared to Sural nerve in lower limbs.

EMG: (Table 2)

EMG: EMG studies were carried out in 30 of the 65 patients of the GBS group, of which 25 (83.33%) showed demyelinating type of polyneuropathy with reduced voluntary motor unit recruitment while the remaining 5 (16.66%) patients showed axonal type of polyneuropathy showing denervation pattern

Discussion

- Electro-diagnostic studies are helpful in diagnosis and differentiating demyelinating variety of GBS which responds to treatment

and has a better prognosis¹³. Electrophysiological hallmarks of early demyelination include prolonged distal motor latencies, prolonged/absent F wave latencies mainly in the lower limbs, slow motor conduction velocities/conduction block with absent F wave, and abnormal upper extremity sensory nerve action potential as compared to the sural nerve.

➤ F wave is the most sensitive diagnostic test for early GBS. In our study, motor conduction velocity was decreased and proximal conduction block was noticed mainly in the lower limbs. The above results are in tandem with findings of Gordon, Kimura & Kuwahara^{4, 10, 14}. In a study done by Ropper et al on 41 patients of GBS who underwent electro-diagnostic studies within a week of onset of symptoms, 16 patients had abnormalities of compound muscle action potentials including dispersion,

delayed latency, low amplitude, conduction velocity slowing, conduction block or abnormal F-waves¹⁵. Similar results have been quoted by Clouston et al¹⁶

➤ Prolonged distal motor latencies & prolonged or absent F waves reflect early predilection for involvement of proximal spinal roots and distal motor terminals. Upper limb SNAP's particularly of the median nerve can be affected more severely and earlier than those of the sural nerve. The explanation of this finding is multifactorial. Entrapment sites are more prone i.e. median nerve in Carpal Tunnel. Reduced SNAP amplitudes can be the result of secondary axonal degeneration and conduction block¹⁷. The conduction block was maximal in the terminal segment in the upper and lower limbs, more so in the lower limb. These findings were consistent with those of Brown¹⁸. He attributed this maximal block

to the relative deficiency of the blood nerve barrier. Decrease in conduction velocity is damage to the myelin sheath; both cellular and immune mechanisms play important roles in it. Early inflammatory lesions consist of lymphocytic infiltrate; later on macrophages become prominent. The peripheral nerve changes consist of perivascular oedema, accumulation of mono-nuclear cells and para-nodal & less commonly segmental demyelination¹⁹.

➤ The EMG studies were conducted on 30 of the 65 patients because of proximity of electro-diagnosis to the symptom onset. EMG studies showed that demyelinating type of neuropathy was the predominant form of GBS (83.33%) in our series. This was in consistence with the results of Yakoob et al²⁰.

Conclusion:

The global incidence of Guillain Barre Syndrome has been estimated between 1.1/100,000/year to 1.8/100,000/year.²¹ Thus; this syndrome constitutes a major load of demyelinating polyneuropathy cases worldwide. There seems to be a slight preponderance of AIDP (acute inflammatory demyelinating polyneuropathy) variety in India as suggested by Gupta et al and Meena et al^{22, 23}. AIDP was the preponderant variant in our study i.e. in the state of Haryana. The results were in line with the electro-diagnostic criteria for early diagnosis of GBS as described in literature. Electro-diagnostic techniques play an important role in the early detection and characterization of inflammatory demyelinating polyradiculopathy in the first week of symptomology and assume importance in treatment of this syndrome because timely intervention reduces morbidity and disability

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Conflict of interest: No conflict of interests

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Table 2: Electro-diagnostic findings in patients with Guillain Barre Syndrome within 1 week

Variable	No (%) of patients
1. F waves	
Abnormal, total	
a. Upper limb	
Absent	50 (76.92%)
Prolonged latency	15 (23.07%)
Normal	0
b. Lower limb	
Absent	50 (76.92%)
Prolonged latency	15 (23.07%)
Normal	0
2. SCV	
a. Upper limb	
Normal	55 (84.61%)
Decrease	05 (7.69%)
Absent	05 (7.69%)
b. Lower Limb (Sural)	
Normal	40 (61.29%)
Decrease	20 (30.76%)
Absent	5 (7.69%)
3. CMAP (compound muscle action potential)	
A. Conduction velocity (CV) & amplitude	
a. Upper limb	
Decrease in both Ulnar & median nerves	44 (67.6%)
Decrease in only ulnar nerve	8 (12.3%)
Normal CV	13 (20.1%)
b. Lower limb	
Decrease in both tibial & peroneal nerves	50 (76.92%)
Decrease in only tibial nerve	5 (7.69%)
Conduction block	10 (15.38%)
B. Distal motor latency	
a. Upper limb	
Increase in both Ulnar & median nerves	42 (64%)
Increase in only ulnar nerve	8 (12%)
b. Lower limb	
Increase in both tibial & peroneal nerves	50 (76.92%)
Increase in only tibial nerve	5 (7.69%)
Conduction block	10 (15.38%)
4. EMG	
Demyelinating type	60 (92.30%)
Axonal type	5 (7.69%)