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ORIGINAL ARTICLE

Clinical and electrophysiological features of Guillain-Barré syndrome: A 7-year experience of Hacettepe University Department of Neurology

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ABSTRACT

Objective: Guillain-Barré Syndrome (GBS) is the most common cause of severe acute paralytic neuropathy. In this retrospective study, we present the GBS experience of the Hacettepe University Department of Neurology and discuss the clinical and electrophysiological features.

Material and Methods: Demographic data, presenting complaints, cerebrospinal fluid (CSF) findings, EMG results, and treatments of patients with electrophysiological findings compatible with GBS between August 2017 and August 2024 were retrospectively reviewed.

Results: Fifty-nine patients diagnosed with GBS (41 males, mean [min-max] age 49 [3–92] years) were recorded. Among them, 66% (n=39) were classified as AIDP, 22% (n=13) as AMAN, 3% (n=2) as AMSAN, and 9% (n=5) as MFS. It was noted that EMG was performed within an average of 11 days after the onset of the first symptom. Sensory complaints were the initial symptoms in half of the patients. Six patients (10%) required mechanical ventilation during follow-up. Prolonged or absent F-waves in one or more nerves were recorded in 64% (n=38) of upper limbs and %90 (n=53) of lower limbs. Reduced distal compound muscle action potentials (CMAP) in two or more nerves were observed in 51 patients (86%), prolonged distal motor latency (DML) in two or more nerves in 40 patients (68%) and slowed motor nerve conduction velocities (NCV) in two or more nerves in 26 patients (44%). Sural SNAP amplitudes were normal in 33 patients (56%), decreased in 14 (24%), and absent in 12 (20%). The conduction block was recorded in 10 patients (17%), and temporal dispersion was noted in 19 patients (32%). Treatment included IVIg in 40 patients (68%) and plasmapheresis in 2 (3%), and due to insufficient treatment response, the number of patients requiring plasmapheresis after IVIg or vice versa was 13 (22%).

Conclusion: This study showed that AIDP is the most common electrophysiological subtype of GBS in our population. Our study found that AIDP was GBS's most common electrophysiological subtype. Prolonged or absent F-waves reduced distal CMAP amplitudes, prolonged DML, slowed NCV, decreased upper limb SNAP amplitudes, and partially preserved sural sensory responses observed in our study were consistent with the findings of other studies.

Keywords: Guillain-Barré Syndrome, electromyography, neuromuscular disease, immunoglobulins, intravenous, plasmapheresis.

INTRODUCTION

Guillain-Barré Syndrome (GBS) is an immune-mediated disease characterized by acute peripheral neuropathy and is a significant cause of neurological morbidity worldwide [1]. Its pathogenesis involves cellular and humoral immunity. The clinical spectrum of GBS includes subtypes such as acute inflammatory demyelinating polyneuropathy (AIDP), acute motor-sensory axonal neuropathy (AMSAN), acute motor axonal neuropathy (AMAN), and Miller-Fisher Syndrome (MFS) [2]. AIDP is the most common form in Western countries, while axonal forms such as

AMAN and AMSAN are more prevalent in Asia and South America.

Although the exact etiology of GBS remains unclear, preceding viral or bacterial infections are believed to trigger antigenic responses in the peripheral nerves [3]. The diagnosis of GBS is based on clinical history, neurological examination, and electrophysiological studies. Key features include the absence of F-waves, prolonged distal motor latency, and sensory nerve action potential (SNAP) abnormalities, which are crucial for early diagnosis [4]. While SNAP amplitudes in the upper extremities were reduced, the preserved sural SNAP pattern has been reported in the literature to increase diagnostic sensitivity. Intravenous immunoglobulin (IVIg) and plasma exchange are effective treatment modalities that prevent mortality and morbidity in GBS [5].

This study retrospectively reviews the clinical and electrophysiological features of patients diagnosed with GBS in the Hacettepe University Department of Neurology.

METHODS

We searched the medical history of the patients in our EMG laboratory's database, which are available between August 2017 and August 2024, using the keywords "demyelinating,""Guillan Barrè Syndrome," "acute motor axonal neuropathy," "AMAN," "acute motor-sensory axonal neuropathy," "AMSAN," "Miller-Fisher Syndrome" and "MFS". The inclusion criteria were, a confirmed diagnosis of GBS based on clinical, neurological, and electrophysiological evaluations and the availability of complete electrophysiological records. All clinical and electrophysiological data were retrospectively collected from 59 patients diagnosed with GBS.

Demographic characteristics (age, sex), presenting symptoms, the time interval between symptom onset and hospital admission, history of predisposing factors such as surgery or infection (upper respiratory tract infection [URTI], acute gastroenteritis, vaccination, treatments administered, ventilator requirements, cerebrospinal fluid (CSF) analysis, and EMG findings were recorded.

Electrophysiological examinations included distal motor latency (DML), compound muscle action potential (CMAP) amplitude, sensory nerve action potential (SNAP) amplitude, motor and sensory nerve conduction velocities, F-wave latency, and H-reflex response.

This study was designed retrospectively, and permission and consent were obtained to review patient files. Ethical approval was granted by the Ethics Committee of Hacettepe University Faculty of Medicine (Decision No: 2024/10-64).

RESULTS

Between August 2017 and August 2024 were retrospectively reviewed, 59 patients (41 males, mean [range] age 49 [3–92] years) diagnosed with GBS were recorded in our unit. The demographic and clinical features of all patients are summarized in Table 1. Among these patients, 66% (n=39) were

Table 1. Demographic, Clinical, and Electrophysiological Features of Patients Diagnosed with GBS

Parameter	Findings
Age (Median, years)	49 [3–92]
Gender, n (%)	
Female	18 (31%)
Male	41 (69%)
Male/Female ratio	2.2
History of infection, n (%):	
URTI	17 (29%)
Acute gastroenteritis	12 (20%)
Vaccination	1 (2%)
Surgery	1 (2%)
Unknown	28 (47%)
GBS Subtypes, n (%):	
AIDP	39 (66%)
AMAN	13 (22%)
AMSAN	2 (3%)
MFS	5 (9%)
Treatment, n (%):	
IVIg	40 (69%)
Plasmapheresis	2 (3%)
IVIg + Plasmapheresis (or vice versa)	
(Due to insufficient treatment)	13 (22%)
No treatment	2 (3%)

URTI: Upper respiratory tract infection, **AGE**: Acute gastroenteritis, **AIDP**: Acute inflammatory demyelinating polyneuropathy, **AMSAN**: Acute motor-sensory axonal neuropathy, **AMAN**: Acute motor axonal neuropathy, **MFS**: Miller-Fisher Syndrome, **IVIg**: Intravenous immunoglobulin

classified as AIDP, 22% (n=13) as AMAN, 3% (n=2) as AMSAN, and 9% (n=5) as MFS.

Sensory complaints were the initial symptoms in 50% (n=30) of the patients, followed by motor complaints in 47% (n=28) and cranial neuropathy in 5% (n=3). Some patients had multiple symptoms simultaneously. A history of infection was noted in 49% (n=29) of the patients within the preceding 30 days. Among these, 17 (59%) had URTI, 12 (41%) had acute gastroenteritis, and one patient had undergone lumbar spinal stenosis surgery two weeks before symptom onset. In 47% (n=28) of the patients, no predisposing factors were identified before GBS symptoms.

CSF sampling was performed within an average of 12 (range:2-45) days after symptom onset, revealing elevated protein levels with a mean of 125 mg/dL (range: 25-354 mg/dL). CSF protein levels of 20 patients could not be obtained for various reasons, such as the absence of CSF sampling or the patient being referred to our center from an external hospital. Among the 39 patients who underwent CSF sampling, 29 (74%) had elevated CSF protein levels (N:15-40 mg/dL).

Regarding treatment, 69% (n=40) of the patients received IVIg, while 3% (n=2) underwent plasmapheresis. Due to insufficient treatment response, 22% (n=13) required plasmapheresis after IVIg or vice versa. Additionally, 3% (n=2) underwent physical therapy and rehabilitation. Two patients were transferred due to hospital bed availability issues, and no data on their treatment course was available. During follow-up, mechanical ventilation was required in 10% (n=6) of the patients.

EMG was conducted within an average of 11 days after the onset of the first symptom. On average, each patient underwent EMG twice (1–5 times). Prolonged or absent F-wave latencies in one or more nerves were recorded in 64% (n=38) of upper limbs and 90% (n=53) of lower limbs. Reduced distal CMAP amplitudes in two or more nerves were observed in 51 patients (86%), prolonged DML in two or more nerves in 40 patients (68%), and slowed motor nerve conduction velocities in two or more nerves in 26 patients (44%).

In the upper limbs, sensory nerve action potential (SNAP) amplitude reductions in one or more nerves were detected in 26 patients (44%). Sural SNAP amplitudes were normal in 33 patients (56%), decreased in 14 (24%), and absent in 12 (20%). Preservation of sural SNAP in the presence of upper limb SNAP abnormalities was observed in 16 patients (27%).

Table 2. Electrophysiological Findings

Parameter	Findings	Count (n)	Percentage (%)
NCS and Needle EMG	NCS + Needle EMG	45	76.3
	NCS only	14	23.7
F-Wave (Upper Limb)	Prolonged/absent in ≥1 nerve	38	64.4
F-Wave (Lower Limb)	Prolonged/absent in ≥1 nerve	53	89.8
Distal CMAP Amplitude	Reduced in ≥2 nerves	51	86.4
	Reduced in 1 nerve	2	3.3
	Normal	6	10.3
DML	Prolonged in ≥2 nerves	40	67.7
Conduction Block		10	16.9
Temporal Dispersion		19	32.2
Preserved Sural SNAP		16	27.1
Nerve Conduction Velocity	Slowed in 1 nerve	14	23.7
	Slowed in ≥2 nerves	26	44.1
	Normal	19	32.2
H reflex	Number of patients studied	8	13.5
	Absent H reflex*	7	87.5
	Sural DSAP is normal*	6	75

NCS: Nerve Conduction Studies, **CMAP:** Compound Muscle Action Potential, **DML:** Distal Motor Latency, **SNAP:** Sensory Nerve Action Potential

* This value is proportional to the number of patients in whom the H reflex was studied.

Conduction block was detected in 10 patients (17%), and temporal dispersion was noted in 19 patients (32%). H-reflexes were evaluated in eight patients and were absent in seven (88%). Among these seven patients, six had normal sural SNAP amplitudes. All electrophysiological findings are summarized in Table 2.

DISCUSSION

We reviewed the data of 59 patients diagnosed with GBS in our center over 7 years. We compared the clinical and electrophysiological data we obtained with the features reported in the literature.

Guillain-Barré Syndrome has an annual incidence of approximately 1.3 cases per 100,000 population globally and is more commonly observed in males than females [6]. Although GBS can occur at any age, its frequency increases in young adults and individuals over 55. Our study similarly demonstrated a male predominance (n=41, 70%) and a median age of 49 years [3–92], consistent with the literature.

Classical GBS symptoms often begin with weakness, occasionally accompanied by paresthesia. The weakness is relatively symmetrical, affecting both distal and proximal muscles of the lower extremities, and frequently progresses to the upper extremities within days or even hours. Weakness progresses rapidly, with 50% of patients reaching maximum disability within two weeks and 90% within four weeks [7]. Reflex loss is a common early finding. The facial nerve is most involved among cranial nerves, with facial diplegia occurring in approximately half of the patients. Ocular motor dysfunction is observed in 15% of cases, with complete ophthalmoplegia in 3–5%. In our study, the most common presenting symptoms were sensory complaints (30 patients, 50%), followed by motor symptoms (28 patients, 47%), cranial nerve involvement in three patients (5%), and autonomic symptoms in one patient (2%). Some patients had multiple presenting symptoms simultaneously.

Between 50% and 70% of GBS cases are associated with infections such as gastroenteritis, respiratory tract infections, or vaccinations within 2–4 weeks before symptom onset [7]. Campylobacter jejuni is most frequently linked with the axonal form of GBS, while cytomegalovirus infections are more often associated with sensory symptoms and cranial nerve involvement [8]. Rarely, GBS may occur following surgery or as part of other diseases [9]. In our study, 49% of patients (n=29) reported a history of infection within the 30 days preceding symptom onset, including 17 cases of upper respiratory tract infections (59%) and 12 cases of acute gastroenteritis (41%).

GBS progresses rapidly, with half of the patients reaching maximum disability within two weeks and 90% within four weeks. More than half of affected individuals lose the ability to walk, and approximately one-third of hospitalized patients require respiratory support and intensive care unit monitoring [10]. In our study, 10% of patients (n=6) required mechanical ventilation during follow-up.

Diagnostic support for GBS includes CSF examination and electrophysiological studies. In CSF, protein levels are elevated, and the cell count is typically $<10/mm^3$. In our study, EMG was performed within an average of 11 days after the onset of symptoms, and CSF sampling was conducted at a mean of 12 days. The mean CSF protein level was 125 mg/dL.

Electrophysiological studies, particularly nerve conduction studies (NCS) and needle EMG, play a vital role in confirming the clinical diagnosis of GBS. Accurately identifying the electrophysiological subtypes of GBS is essential for understanding the disease's dynamic nature. Sequential electrophysiological evaluations are key in achieving this and ensuring precise subtype classification.

Uncini's criteria aim to minimize diagnostic errors by providing detailed definitions of these dynamic processes and the specific features of different subtypes [12]. In most of our patients (n=36,61%), EMG was performed at least twice. The most well-known form, AIDP, is observed in 80–95% of cases, particularly in Europe and North America [11]. While GBS is often used interchangeably with AIDP, other forms exist, such as AMAN and AMSAN. In

our study, AIDP was the most frequently observed subtype (66%, n=39), consistent with the literature.

Diagnosis of AIDP is based on findings such as motor nerve conduction velocities <70%, distal motor latencies >130% of normal values, distal CMAP durations >120%, and F-wave latency >120% in at least two nerves. If only one of these findings is present, the absence of F-waves in two nerves and decreased ulnar SNAP amplitudes with preserved sural SNAP amplitudes should be noted. Axonal subtypes (AMAN and AMSAN) are characterized by the absence of AIDP-specific findings, with features such as distal CMAP amplitudes reduced to <80% and conduction block in at least two nerves [12]. AMSAN is characterized by significant damage to sensory nerves, with at least a 50% reduction in SNAP amplitudes in at least two nerves, persisting in subsequent examinations. Additionally, acute denervation signs, such as fibrillation potentials and positive sharp waves, typically appear 7–10 days after symptom onset [13]. In our study, patients underwent an average of two EMG studies (range: 1–5). Findings such as prolonged or absent F-waves, reduced distal CMAP amplitudes, prolonged distal motor latencies, and preserved sural SNAP amplitudes observed in our study were consistent with previous literature.

The treatment of GBS relies on a multidisciplinary approach, with immunotherapy and comprehensive supportive care serving as the foundation of effective management. Effective treatment modalities include IVIg and plasma exchange. In our clinic, IVIg was the most frequently administered treatment.

CONCLUSION

Our study findings demonstrate that the clinical and electrophysiological features of GBS observed in our cohort align with existing literature. The acute phase of GBS, characterized by rapid progression, underscores the importance of early diagnosis and close monitoring. Electrophysiological evaluations and serial follow-up studies play a critical role in guiding treatment and providing valuable insights.

Ethical approval

This study was designed retrospectively, and permission and consent were obtained to review patient files. Ethical approval was granted by the Ethics Committee of Hacettepe University Faculty of Medicine (Decision No: 2024/10-64).

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Conflict of interest

The authors declare that there is no conflict of interest.

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