

Study of clinical profile and prognosis in various subtypes of guillain barre syndrome patients

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Abstract

Guillain-Barré syndrome is a post infective polyradiculoneuropathy having heterogenous clinical presentation and various subtypes like acute inflammatory demyelinating polyradiculoneuropathy, acute motor-sensory axonal neuropathy, acute motor axonal neuropathy, pure sensory variant and Miller Fisher syndrome. A prospective study was carried out in patients with Guillain-Barré syndrome admitted to the RNT Medical College & attached Hospital to determine the electrophysiological subtypes and their prognosis in relation to various subtypes, clinical features and treatment. A total of 100 patients was enrolled. In the final analysis there were 74% male and the mean age was 30.4 years. Clinically 97% patients had quadriplegia, 2% had paraparesis and one cases had bibrachial involvement. Cranial nerves and respiratory involvement were seen in 25% and 24% cases respectively. Electrophysiologically the most common type of GBS was AIDP (43%) followed by AMAN (34%) and AMSAN (23%). The prognosis was assessed at one month and found that there was complete recovery in 32% cases and residual weakness in 63% cases. Death occurred in 5% cases because of respiratory involvement.

Introduction

Guillain Barre Syndrome (GBS) is a disorder of peripheral nerves and roots and sometime cranial nerves which is considered to be an autoimmune disease triggered by a preceding bacterial or viral infection and it is mostly characterized by an acute onset ascending weakness of more than two limbs progresses over a period of days and up to 4 weeks, hypo or areflexia and albuminocytological dissociation in cerebrospinal fluid examination. The incidence of GBS is almost same throughout the year with a rate of 1 to 4 cases per 100,000 annually and in the United States, 5000–6000 cases occur per year.¹ It can occur in any age group but in western countries adults are more frequently affected than children.¹ Males are at slightly higher risk for GBS than females, Seasonal variations are noted in some series. The classification of GBS is based on electrophysiological study into acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor-sensory axonal neuropathy (AMSAN) and acute motor axonal neuropathy (AMAN), pure sensory variant and Miller Fisher syndrome (MFS).^{2,3} We have studied the clinical, electrophysiological parameters and their prognosis in regards to their presentation and treatment.

Aims and Objectives

1. To analyses the clinical profile of GBS.
2. To analyses the electrophysiological changes and subclassified in various types at time of admission.
3. To study the prognosis in GBS in various subtype.

Materials and Methods

100 patients of Guillain Barre Syndrome (GBS) who were diagnosed by clinical and

electrophysiological study and supported by CSF findings admitted in the of R.N.T. Medical College and Associated Hospital, Udaipur from 1st January 2015 to 30th June 2018 were included in this study.

A detailed history and clinical examination was performed at the time of admission and repeat examinations of muscle power were performed on every day till discharge.

During each examination the muscle power was graded according to Medical Research Council (MRC).

Routine blood investigations including complete blood count, Renal; function test, liver function tests were performed all patients were also screened for hepatitis B, C serology and HIV serology. Cerebrospinal fluid examination was done in all patients to see cell count and protein, sugar levels to support the diagnosis.

Electrophysiological study was performed in all patients. With proper guideline and protocols. Limb temperature was maintained above 32° C using warmer, if needed. Motor and sensory nerves conduction studies were performed in at least four motor nerves (Median, ulnar, tibial, and peroneal) and three sensory nerves (Median, ulnar, and sural). In motor nerves, distal latency, amplitude and nerve conduction velocity (NCV), were evaluated.⁴ F-wave minimal latency and evidence of conduction block were also. In sensory nerves peak latency, velocity and amplitude were measured.

The cases were finally classified into three groups: AIDP, AMAN, or AMSAN. AIDP was diagnosed based on Albers and Kelly criteria.⁵ the patients with no evidence of demyelination, they were classified into AMAN and AMSAN whether there is sensory involvement present or not.

Electrophysiological criteria for classification of Guillain-Barré syndrome (adapted from Ho et al⁶)

A. Diagnosis of demyelination: Present in two or more nerves

1. Conduction velocity <90% of lower limit normal if amplitude is >50% of lower limit normal; <85% if amplitude <50% of lower limit of normal
2. distal latency >110% of upper limit of normal if amplitude normal ;>120% of upper limit of normal, if amplitude is less than lower limit of normal
3. Evidence of unequivocal temporal dispersion
4. F-latency >120% of normal

B. Diagnosis of primary axonopathy:

1. No evidence of demyelination as above
2. Decrease in CMAP (compound muscle action potential) to <80% of lower limit of normal
3. Denervation changes on needle electromyography

Inclusion Criteria

1. Age more than 2 years
2. Patients who are diagnosed as Guillain Barre Syndrome as per criteria proposed by Asbury and Cornblath⁵ in 1990

Exclusion criteria

All patients who were having chronic medical disease and other underlying cause of neuropathy like acute or chronic renal failure, cirrhosis of liver, meningitis and head trauma Tuberculosis, asthma, Cystic fibrosis, certain drugs like SSRI, TCA, Diuretics, Nicotine, Narcotics and patients with malignant diseases were excluded from study.

All patients of GBS were hospitalized and based on severity they were treated conservatively and with mechanical ventilatory assistance, where required. Most of patients were treated with intravenous Immunoglobulin (IVIG, 0.4g/kg/for 5 days) and some patients treated with intravenous methylprednisolone for 5 days where there is nonavailability of IVIG and patients having no contraindication to steroids.

Observations and Results

This study has been conducted on 100 patients of GBS. There were 74% male and the mean age was 30.4 years.

Clinically 97% patients had quadriparesis, 2% had paraparesis and one cases had bibrachial involvement. Cranial nerves and respiratory involvement were seen in 25% and 24% cases respectively. Sensory involvement was seen in 14% cases.

Electrophysiologically in our study the AIDP (43%) was most common followed by AMAN (34%) and AMSAN (23%). In AIDP 16 out of 43 cases had sensory involvement. There was no significant seasonal clustering among electrophysiological subtypes.

The prognosis was assessed at one month and found that there was complete recovery in 32% cases and residual weakness in 63% cases. Death occurred in 5% cases because of respiratory involvement.

The other prognostic parameters like age and sex of patients, days of symptoms onset to presentation to hospital, clinical involvement, electrophysiological findings and treatments options were assessed and shown in figures and tables below.

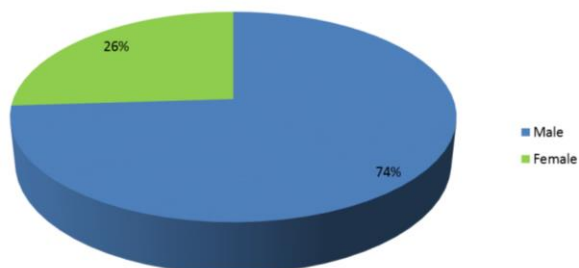


Fig. 1: Sex Ratio of study subjects

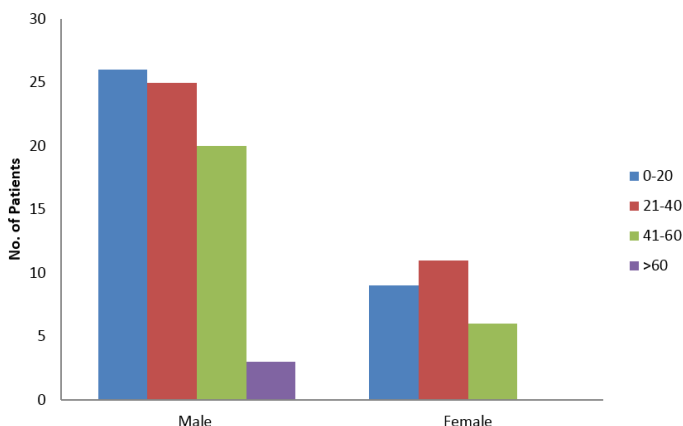


Fig. 2: Age Group of study subjects

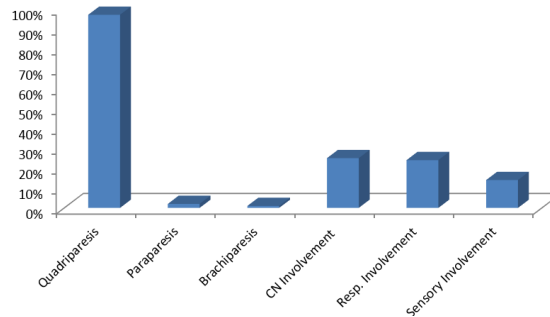


Fig. 3: Clinical Presentation of study subjects

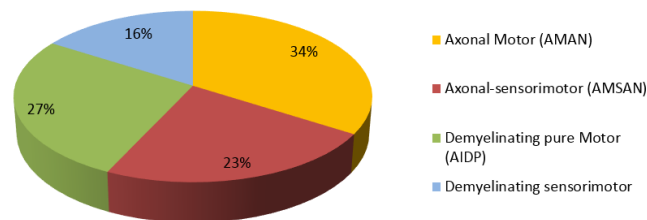


Fig. 4: Subtypes of GBS based on electrophysiological study

Table 1: Outcome of study subjects after 1month follow up:

| Outcome | No. of Patients |
|----------|-----------------|
| Complete | 32% |
| Residual | 63% |
| Death | 05% |
| Total | 100 |

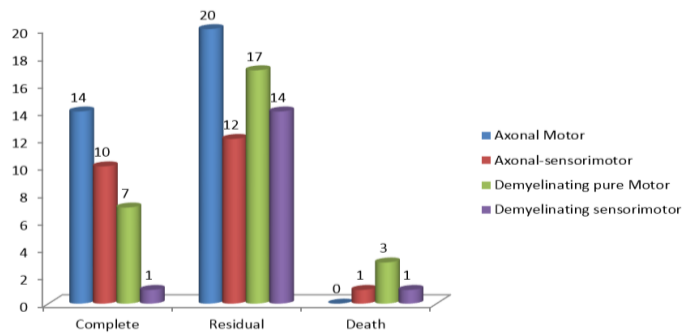


Fig. 5: Prognosis According to Nerve Conduction Study

Table 2: Days of Illness

| Outcome | <7days | 8-14 days | >14 days |
|----------|--------|-----------|----------|
| Complete | 23 | 2 | 7 |
| Residual | 45 | 3 | 15 |
| Death | 5 | 0 | 0 |
| Total | 73 | 5 | 22 |

Table 3: Prognosis according to age

| Outcome \ Age Group | 0-20 | 21-40 | 41-60 | >60 | Total |
|---------------------|------|-------|-------|-----|-------|
| Complete | 19 | 11 | 2 | 0 | 32 |
| Residual | 14 | 24 | 22 | 3 | 63 |
| Death | 2 | 1 | 2 | 0 | 5 |
| Total | 35 | 36 | 26 | 3 | 100 |

Table 4: Prognosis according to Respiratory involvement

| Outcome \ Resp.Inv. | Yes | No | Total |
|---------------------|-----|----|-------|
| Complete | 2 | 30 | 32 |
| Residual | 17 | 46 | 63 |
| Death | 5 | 0 | 5 |
| Total | 24 | 76 | 100 |

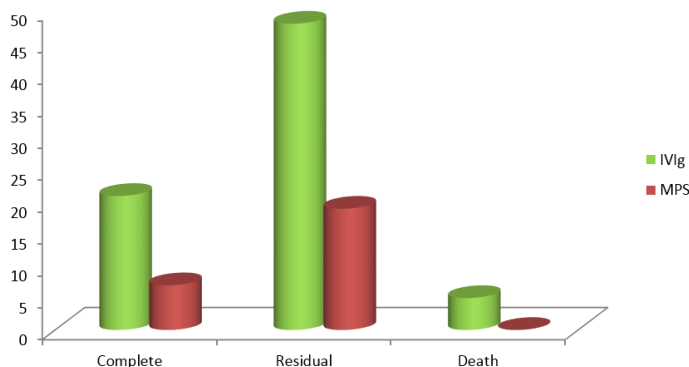


Fig. 6: Prognosis according to Treatment given

Discussion

There are many studies in patients of Guillain barre syndrome and its subtypes regarding its clinical presentations and prognosis. Still many times we come across various atypical presentations like asymmetrical involvement and variable sensory symptom and signs. The diagnosis of Guillain-Barré syndrome was based on clinical features,⁷ with evidence of demyelination on electrophysiology supported by CSF findings.^{8,9} However recently it has become evident that the syndrome is heterogeneous with various subtypes, including predominantly axonal patterns; a sensorimotor axonal neuropathy now called acute motor-sensory axonal neuropathy (AMSAN)⁶ and an acute motor axonal neuropathy (AMAN).^{2,3} In our study we found male preponderance with no seasonal variation.

We have male: female ratio 2.8:1 which is comparable to studies done by Maneesh et al¹⁰ (male: female 2.3:1), Gang Zhang et al¹¹ (1.83:1), Hemant bhagat et.al¹² (3:1). In present study 100% cases presented with some motor weakness out of which 97% cases had quadriplegia, 2% had paraparesis and only one case had brachiparesis. The pattern and extent of involvement of motor weakness depends on geographical variation and patients’ immune status. Kalita et al¹³ found in her study that pure motor GBS is more common (62.2% cases) than motor with sensory (32.3%) and concluded that there is one other entity like pure sensory GBS (0.6%). In our study sensory involvement was seen in 14% cases and pure sensory variant was not seen probably because lack of awareness in patients to consult as there is no motor weakness.

In our study cranial nerves (CN), Respiratory and sensory involvement were present in 25%, 24% and

14% cases. Maneesh et al⁷ found that CN, Respiratory and sensory involvement were present in 20%, 15%, and 25% cases respectively. Dhadke et al.¹⁶ found CN involvement in 62.5% cases and sensory involvement in 32.5% cases whereas Lofell et al¹⁷ and Amita Bhargava et al¹⁴ found CN involvement in 50% and 62.5% cases respectively. We did not find any case of Miller fisher syndrome (MFS).

Electrophysiologically maximum number of patients has the AIDP pattern (43%) followed by AMAN (34%) and AMSAN (23%). Mostly AIDP is considered as pure motor illness but in our study 37.20% cases of AIDP had sensory involvement which is a new thing and needs more data in future to comments on variable pattern and more sensory involvement. J kalita et al¹³ from India got same result as our study. They concluded that AIDP (73.8%) was most common form of GBS followed by AMAN (13.4%), and AMSAN (4.6%). Gang zhang et al¹¹ from china found AMAN type was most common and had better prognosis than AIDP. Verma et.al found axonal pattern in 60% and demyelinating pattern in 30% cases. The disparity may be because of geographical variation and study groups population. Our study has more adult patients.

The prognosis of recovery depends on so many factors like age of patients and clinical findings especially respiratory involvement as well as time of presentation to symptoms onset and immunomodulator therapy. Sometime the prognosis also depends on other comorbidities and metabolic derangement. In our study, we found that 32% cases recovered completely and 63% partially at the end of one month .24% cases needed mechanical ventilation, out of which 5 patients died and remaining survived. The death in those cases was due to aspiration and autonomic involvement. We

have given intravenous immunoglobulin to 74% cases, rest (24%) had been treated by high dose Methyl prednisolone (MPS) for 5 days. No one treated by plasmapheresis because of unavailability. In MPS treated groups 26.9% cases improved completely which tells that still MPS is an alternative if not contraindicated as it is easily available and cheaper options and it can modify the immunological status of the patient and help in recovery. Kaul et al¹⁵ found that 11 out of 61 cases in their study needed mechanical ventilation. They also concluded that mechanical ventilation, CN involvement and Axonal variety were poor prognostic factors.

Conclusion

The Guillain barre syndrome still has variable presentation in terms of severity and symmetry of limb involvement and cranial nerves involvement. The prognosis of recovery and death depends on many factors like involvement of respiratory muscles, autonomic nervous system as well as treatment options i.e. IVIG or plasmapheresis. There is still a hope for methylprednisolone where not contraindicated and unavailability of plasmapheresis or immunoglobulin.

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