

Clinical Profile, Electrodiagnostic Variations, Treatment and Outcome in Guillain-Barré Syndrome: Prospective Study of 78 Patients

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ABSTRACT

Introduction: Guillain-Barre syndrome (GBS) is an acute polyradiculoneuropathy with varied severity of presentation. Objectives: To study clinical presentation, electrodiagnostic variations, hospital care and outcome of patients of Guillain Barre Syndrome (GBS). Current research aimed to study efficacy of Plasmapheresis and Intravenous Immunoglobulin (IVIG) in patients of GBS.

Material and Methods: 78 patients of GBS were studied in detail including history, clinical examination and investigations. Patients were treated with Plasmapheresis and IVIG; and outcome was observed.

Results: Commonest age group affected was 13-40 yrs. The male:female ratio was 1.2:1. Antecedent infection were present in 42 out of 78 patients. Quadripareisis was present in 74 patients and paraparesis in four patients. Cranial nerve involvement was seen in 47 out of 78 patients. Areflexia was found in all 78 patients. Albuminocytologic dissociation was present in 44 out of 61 patients underwent CSF examination. NCV findings show conduction velocity slowing, delayed f latencies in 92% patients. Out of 72 patients, 33[43%] required mechanical ventilation. Out of 78 patients, 71 were treated with Plasmapheresis and seven patients were treated with IVIG. Out of 78 patients 56 [72%] patients recovered completely, 19 [24%] patients died and three [four%] patients developed severe neurologic deficit.

Conclusion: GBS is more common in male as compare to female. Commonest presentation is paresthesia in legs and ascending paralysis. Patients who received treatment early in the course of disease had faster recovery. Plasmapheresis and IVIG both showed similar efficacy in treating GBS.

Keywords: Guillain-Barre Syndrome, Plasmapheresis, IVIG.

INTRODUCTION

Guillain-Barre Syndrom is an acute, frequently severe and fulminant polyradiculoneuropathy that is autoimmune in nature.¹ GBS manifests as rapidly evolving areflexic motor paralysis with or without sensory disturbance. The usual pattern is an ascending flaccid paralysis.² Autonomic involvement is common. Respiratory failure occurred and ventilatory assistance was required in 30% cases.^{1,3,4} 70% cases are post infectious 1-3 weeks after an acute infectious process respiratory or GIT.⁵ 20-30% cases of them are due to campylobacter jejuni. Other agents are HHV (EBV,CMV) and Mycoplasma pneumonia.⁶ Swine flu vaccine and older rabies vaccine are rare causes.^{7,8} The theories suggest an autoimmune mechanism in which the patient's defense system of antibodies and WBC are triggered into damaging the nerve coverings or insulation leading to weakness and abnormal sensation.⁹ To confirm diagnosis nerve conduction

velocity (NCV) is performed.¹⁰ The American Academy of Neurology practice guidelines has recommend either IVIG or plasmapheresis for GBS patients.

Current research aimed to study clinical presentation, Hospital course and outcome in patients of Guillain-Barre syndrome admitted in tertiary care institute, to study the number of patients developing respiratory failure and requiring artificial ventilation and to evaluate the clinical benefits of plasmapheresis and IVIG in patients of Guillain-Barre syndrome.

MATERIAL AND METHODS

A prospective observational study was conducted from June 2017 - July 2018 among the patients admitted in Civil Hospital, Ahmedabad in Gujarat where 78 consecutive GBS patients, underwent detailed clinical and electrophysiological assessment. Institutional Ethics Committee approved the study protocol and written informed consent was taken from all relatives of participant. In this study all male-female patients with age > 13 years with Gullian barre syndrome admitted into our hospital were included and pregnant, lactating woman, patients with significant cardiac, respiratory, renal, psychiatric morbidity and terminal illnesses were excluded from the study.

We did detailed clinical history recording and examinations of each patient. We used Medical Research Council (MRC) sum score for assessing functional motor deficits. For all patients, clinical assessment was done initially at a time of admission, then at four weeks and eight weeks after treatment with plasmapheresis and IVIG. Investigations were done in the form of CBC, peripheral smear examination, renal and liver function test including Serum electrolytes

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How to cite this article: Chirag J Patel, Jigarkumar B Gosai, Divyesh B Kalariya, Manoj Rathod, Surendhar S. Clinical profile, electrodiagnostic variations, treatment and outcome in Guillain-Barré Syndrome: prospective study of 78 patients. International Journal of Contemporary Medical Research 2019;6(8):H1-H6.

DOI: <http://dx.doi.org/10.21276/ijcmr.2019.6.8.42>

Characteristic	Male	Female	Total no of patients
Antecedent event:			
Fever, Cough, Sneezing	11(14%)	07(9%)	18(23%)
Loose motion, Vomiting	15(19%)	09(11%)	24(30%)
Vaccination	00	00	00
Exanthematous fever	00	00	00
Motor system involvement:			
Quadriparesis	43(55%)	31(39%)	74(94%)
Paraplegia	00	04(5%)	04(5%)
Atypical presentation	00	00	00
Areflexia	43(55%)	35(44%)	78(100%)
Cranial nerve involvement:			
Occulomotor	02(2.5%)	01(1.28%)	03(3.84%)
Facial (Unilateral)	01(1.28%)	01(1.28%)	02(2.56%)
Facial (Bilateral)	12(15.38%)	07(9%)	19(24.35%)
Glossopharyngeal, Vagus	17(21.79%)	06(7.69%)	23(29.48%)
Spinal Accessory	00	00	00
Hypoglossal	00	00	00
Autonomic dysfunction:			
Arrhythmia	07(9%)	02(2.5%)	09(11.53%)
Fluctuation in pulse	09(11.53%)	03(3.84%)	12(15.38%)
Fluctuation in BP	14(18%)	07(9%)	21(27%)
Sweating abnormalities	18(23%)	08(10%)	26(33%)
GI dysfunction	04(5.12%)	02(2.5%)	06(7.69%)
Postural hypotension	03(3.84%)	09(11.53%)	12(15%)
Pupillary abnormalities	03(3.84%)	01(1.28%)	04(5.12%)
Urinary retension	00	00	00
Albumino-cytogenic dissociation	25(32%)	19(24%)	44(56%)
NCV studys			
Absent/Delayed f-wave latencies	40(51.28%)	32(41%)	72(92%)
Conduction velocity slowing	40(51.28%)	32(41%)	72(92%)
Prolonged distal motor latencies	31(39.74%)	35(44.30%)	66(84.61%)
Reduced CMAP amplitude	28(35.89%)	37(47%)	65(83%)
Absent/↓ SNAPs	7(9%)	11(14.10%)	18(23%)
Electrodiagnostic categorization:			
AIDP	26(33%)	19(24.35%)	45(57%)
AMAN	07(9%)	05(6.41%)	12(15.38%)
AMSAN	02(2.5%)	04(5%)	06(7.5%)
Unclassified(Mixed)	08(10%)	07(9%)	15(19%)
TREATMENT:			
IVIg	2(2.5%)	5(6.41%)	07(9%)
Plasmapheresis	41(52.56%)	30(38.46%)	71(91%)
Duration of mechanical ventilation:			
0-10 days	08(10.25%)	06(7.5%)	14(18%)
10-20 days	06(7.5%)	03(3.84%)	09(11%)
20-30 days	05(6.41%)	02(2.5%)	07(9%)
>30 days	02(2.5%)	01(1.28%)	03(3.84%)
Duration of hospital stay			
0-10 days	05(6.41%)	03(3.84%)	08(10.25%)
10-20 days	23(29.48%)	11(14.10%)	34(43.58%)
20-30 days	11(14.10%)	15(19.23%)	26(33.33%)
>30 days	04(5.12%)	06(7.69%)	10(12.82%)
Outcome:			
Complete recovery	29(37.17%)	27(34.61%)	56(71.79%)
Incomplete recovery	02(2.5%)	01(1.28%)	03(3.84%)
Death	12(15.38%)	07(9%)	19(24.35%)

Table-1: Characteristics of patients included in the study:

Characteristics	Amita Bhargav et al ¹²	Sudulugunta SR et al ¹³	Maneesh Kumar et al ¹⁴	Dhadke SV et al ¹¹	Ujjal KR Sarkar et al ¹⁵	Kalita J et al ¹⁶	Sandha Manoj et al ¹⁷	Present study
1 Type of the study	Prospective	Retrospective	Cross sectional	Prospective	Retrospective	Cross sectional	Prospective	Prospective
Study population (n)	61	1166	20	40	139	51	36	78
Study period (Months)	26	132	18	20	120	-	48	13
Age range(Yrs)	8-78	0-85	1.3-17	13 -40	2-12	-	19-68	13-66
Male: Female ratio	2.5:1	1.07:1	2.3:1	1.5:1	2.23:1	-	1.4 : 1	1.2:1
Preceding events prior to the illness (%)	47.5	89.7	50	55	65.47	-	63.8	53
3 Clinical Presentation(%)								
Limb weakness	98	99.8	100	100	100	-	97.2	100
Sensory dysfunction	70	37.82	25	32.5	58.9	-	80.5	23.07
Facial palsy	45.9	34.39	-	30	-	-	61.1	27
Autonomic dysfunction	27	-	20	-	35.25	-	36.1	39.74
Respiratory distress	-	16.98	35	32.5	-	-	27.7	42.3
4 Albumino-cytological dissociation in CSF analysis (%)	80	-	-	65.3	38.85	-	81.8	56.41
5 Electro diagnostic categorization(%)								
AIDP	60.6	-	22.2	100	89.2	86.3	34.2	45
AMAN	32.8	-	38.9	-	-	7.8	25.7	15
AMSAN	5	-	22.2	-	-	5.9	25.7	08
Unclassified/Mixed	-	-	16.7	-	-	-	11.4	19
6 Recipients of IVIG(n)	42	51	14	14	139	-	32	7
7 Recipients of plasmapheresis(n)	-	48	1	4	-	-	1	71
8 Outcome(%)								
Complete recovery	75	-	70	75	-	22.4	97.2	56
Incomplete recovery	25	-	15	5	-	57.1	2.77	4

Table-2: Comparison of previous clinical studies with present study:

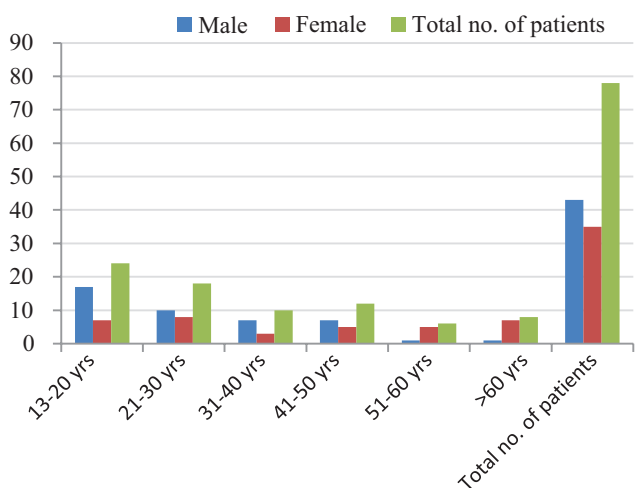


Figure-1: Age and sex distribution

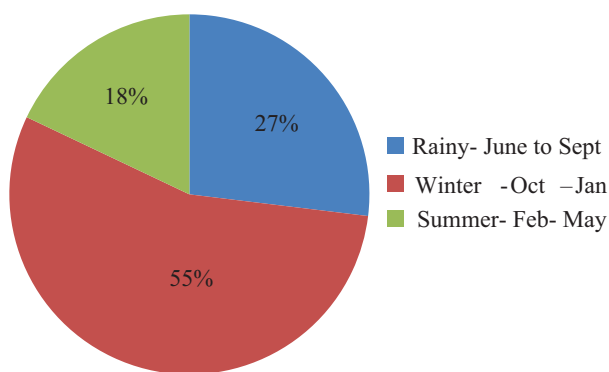


Figure-2: Distribution of the patients according to season:

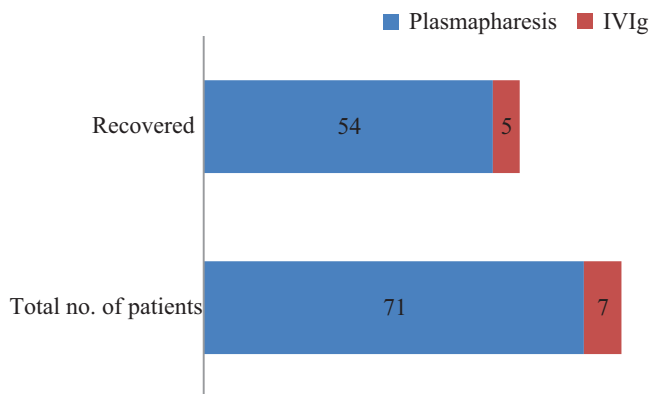


Figure-3: Comparison of outcome in patients treated with Plasmapheresis and IVIg:

in all patients. CSF examination was done for albumino-cytological dissociation and the diagnosis was confirmed by nerve conduction velocity studies (NCV).

Data obtained in the study were entered into Microsoft excel sheet and categorical variables were summarized as counts (percentage).

RESULTS

The present study was undertaken during the period of June 2017 - July 2018. The study comprises of 78 patients. There were 43 males and 35 Females (M:F=1.2:1) their ages ranged from 13 yrs to 66 yrs. The mean age of patients involved in present study was found to be 34 years and median age was

found to be 30 years. Maximum number of patients i.e. 24 (30%) were between 13-20years of age group. Next common age group was of 21-30 years in which 18 (23%) patients were seen. (Figure 1) Antecedent event in patients of GBS studied in which, forty two patients (53%) had preceding illness. (Table 1) Majority of the patients i.e. 24(57%) had gastrointestinal infection as evidenced by loose motion and vomiting. Respiratory infections was encountered in 18 (44%) patients. (Table 1) Majority of the patient 43(55%) found in Winter season. (Figure 2) Majority i.e. 40 out of 42 (95%) patients developed neurologic illness within 4 weeks of antecedent event, while remaining two patients developed it between 1-3 months of preceding illness.

The most common clinical feature was observed quadripareisis in 74 (94%) patients. The least common clinical feature observed was paraparesis in four (6%) patients. Areflexia observed in all patients. (Table 1)

Involvement of cranial nerves in which commonest cranial nerve involved was facial in 21 [45%] patients. Bilateral Facial nerve involvement was observed in 19 patients. Glossopharyngeal and vagus nerve was involved in 23 [48%] patients, while Hypoglossal and spinal accessory nerve involvement were not seen. Oculomotor nerve involvement was seen in three patients. (Table 1) Autonomic dysfunction observed in 31 (39.74%) patients in the form of Arrhythmia in nine patients, Fluctuation in pulse and BP in 12 and 21 patients respectively, sweating abnormality, GI dysfunction, postural hypotension and pupillary abnormalities in 26, 6, 12 and 4 patients respectively. (Table 1) Out of 78 patients, 33 (43%) had respiratory paralysis during the course of illness. Out of 33 patients 28 [36%] required respiratory support within one week of onset of weakness while five patients required ventilatory support within 1-2 weeks of onset of weakness. (Table 1) CSF abnormalities in the form of Albumino-Cytogenic dissociation observed in 44 (72%) patients out of 61 patients in which CSF examination was done and nerve conduction studies were abnormal in 78 patients (100%). (Table 1) Nerve conduction studies were abnormal in 78 patients (100%). Out of 78 patients, 72 (92%) patients had absent / delayed f-wave latencies and slowing of conduction velocity, which was the most common abnormality observed. (Table 1) Majority of the patients 45(57%) were Electrodiagnostic categorized as AIDP while AMAN and AMSAN variety observed in 12(15%) and 6(8%) patients respectively. Majority of the patient 66(85%) had IJV-DLC, while 5(6%) patients have Femoral-DLC. Majority patient 71 (91%) received treatment in form of plasmapheresis, while 7(9%) patient received IVIg. (Figure 3) Duration of mechanical ventilation in majority patients 14(42%) was less than 10 days, around 16(48%) patient required mechanical ventilation for 10-30 days and around 3(10%) patient required mechanical ventilation for more than a month. Respiratory complications were observed in majority 33(42%) patients, while cardiovascular complications in 21 (27%) patients, nutritional complications in 30(38%) patients, metabolic complications in 23(29%) patients and hematologic complication in form of DVT was seen in 1 patient. Duration

of hospital stay in majority 34(44%) patient was 10 to 20 days while few patients 8(10%) required hospital stay of less than 10 days, some patients 26(33%) required 20 to 30 days of admission and around 10(13%) patient admitted for more than 1 month.(Table 1)

Majority of the patient 72(92%) have Low MRC sum score, after 4 weeks and after 6 weeks there is an improvement in MRC score in 54(75%) patients and 56(77%) patients respectively. Out of 71 patients treated with Plasmapheresis 54(76%) recovered fully while out of 7 patient treated with IVIg 5(72%) patients recovered fully. (Figure 3) It was found that the treatment methods employed in the present study were significantly efficacious in treatment of patients with GBS; where 75.64% (59 out of 78) patients showed complete recovery. Majority of the patient 56(72%) were recovered completely after treatment, while 19 patients (24%) patients were died and incomplete recovery was observed in 3 patients (4%) (Table 2).

DISCUSSION

The study gathered information from 78 GBS patients who were > 13 years of age, admitted in the civil hospital Ahmedabad. The study was an attempt to identify the clinical and epidemiological profile of GBS in adults and compared with previous clinical studies.¹¹⁻¹⁷ (Table 2). Most of the studies were conducted either in children or both adults and children. In this study, 52 (66.6%) were, < 40 years and 26 (33.3%) were aged > 40 years. The mean age was 34 years. Sex ratio showed a slight male preponderance in most of the studies¹¹⁻¹⁷ Preceding events prior to the illness in present study is observed in forty two patients (53%) had preceding illness. (Table 1) In most of previous studies shows antecedent event in >50% patients.^{11-15,17} Majority of the patients >97% in all studies have shown limb weakness.^{11-15,17} As compared to other studies in present study there were more number of patients with autonomic dysfunction (39.74%) and respiratory distress (42.3%). Amita bhargav et al¹² and Sandhya Manoj et al¹⁷ observed albuminocytogenic dissociation in about 80% patients while in our study it was around 50.41%. In majority of the studies more number of the patients have Electrodiagnostic categorisation of AIDP variety.^{11,12,15,16} In our study majority >90% patients were treated with Plasmapheresis while in majority of the previous studies^{11,12,14,15,17} >90% patients with GBS treated with IVIg. In our study complete recovery were observed in about 56% patients while in other studies^{11,12,14,17} >70% patients with GBS were recovered completely. Early arrival (<1 week) to hospital, initiation of treatment in form of plasmapheresis or IVIg and intensive care helped to achieve complete recovery in our study. AMAN and AMSAN form of GBS recovered more slowly than those with AIDP form.

CONCLUSION

Guillain Barre Syndrome is common more in males than females. Commonest age group is between 13-40 yrs of age. History of antecedent infection (Gastrointestinal or respiratory) is seen in about 53% of patients. Commonest

presentation of GBS is paresthesia in legs and ascending paralysis. Facial nerve(bilateral) is commonly involved nerve and all patient had abnormal NCV studies. Areflexia is the most common sign observed in 78 patients (100%). 43% of patients developed respiratory paralysis and needed ventilatory support. Nerve Conduction Velocity study is the most useful investigations in diagnosis and abnormal in all GBS patients. Autonomic dysfunctions observed in one third of the patients. In this study plasmapheresis and IVIG both have almost same efficacy in treating GBS. Out of 78 patients 56(72%) were recovered completely after treatment.

ABBREVIATION

GBS - Guillain Barre syndrome, AIDP - acute inflammatory demyelinating polyradiculoneuropathy, AMAN - Acute motor axonal neuropathy, AMSAN - Acute motor sensory axonal neuropathy, IVIG - intravenous immunoglobulin, NCV - nerve conduction velocity, CSF - cerebrospinal fluid, MRC - Medical research council, GIT- Gastrointestinal tract, HHV- Human herpes virus, EBV- Epstein-barr virus, CMV-Cytomegalo virus, WBC- White blood cell, BP-Blood pressure

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Source of Support: Nil; **Conflict of Interest:** None

Submitted: 02-07-2019; **Accepted:** 02-08-2019; **Published:** 28-08-2019