International Journal of Pharmacology and Clinical Research (IJPCR)
IJPCR |Vol.7 | Issue 4 | Oct - Dec -2023
www.ijpcr.com
ISSN: 2349-5448
DOI : https://doi.org/10.61096/ijpcr.v7.iss4.2023.360-366

Research

Clinical and electrophysio-logical profile and functional outcome of guillian barre syndrome patients

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Published on: 22 Dec 2023

Published by:
DrSriram Publications

Abstract

Background and Objectives: Guillain–Barre syndrome (GBS), also known as Landry's paralysis, is an immune-mediated disorder of nervous system of acute or subacute onset characterized commonly by generalized progressive weakness of arms and legs, limb paresthesias and relative or complete areflexia. The present study aimed at documenting clinical and electrophysiological profile and functional outcome of GBS patients along with seasonal variations.
Methods: An observational, cross-sectional study of 41 patients diagnosed with Guillian Barre syndrome admitted in Medicine wards and Medical Intensive Care Unit of Dept of Medicine, GMC, Aurangabad was conducted from October 2019 to October 2021. The data related to demographics, antecedent illness, muscle power on admission, Hughes score on admission and at discharge, CSF studies, electrophysiology, treatment given and the outcome of patients was studied.
Results: Mean age of subjects was 36.63 ± 19.2 years. Ascending paralysis was the most common presentation. In the study majority of cases (60.97%) had AIDP. In the study population, albumino-cytological dissociation in CSF findings was present in 37 patients out of 41(90.24%). There was statistically significant decrease in Hughes score at d ischarge compared to admission score. In the study 16 subjects required Ventilatory assistance. Among them 10 were discharged and 6 died.
Conclusion: Quadriparesis was the most common presentation among Guillian Barre syndrome patients. The majority of the patients had AIDP(Acute Inflammatory Demyelinating Polyneuropathy) subtype of GBS. There was statistically significant decrease in Hughes score in AIDP patients at discharge as compared to the Hughes score at admission. There was no seasonal variation in the occurrence of GBS.

Keywords: GBS, AIDP, Acute motor axonal neuropathy, Acute motor sensory axonal neuropathy, electrophysiology.

INTRODUCTION

Guillain–Barre syndrome (GBS), also known as Landry's paralysis, is an immune-mediated disorder of nervous system of acute or subacute onset characterized commonly by generalized progressive weakness of arms

and legs, limb paraesthesias and relative or complete areflexia[1]. It is an acute polyneuropathy that usually presents as a mono-phasic paralyzing illness[2]. The incidence of GBS in USA is about 1-4 cases per 1,00,000 anually.[3] The incidence of more typical GBS is from 0.81 – 1.89 per 100000 people per year, affecting more men than women and increases with age[4]. GBS is classified into different subtypes according to clinicalpresentation, electrophysio-logical abnormalities and the presence of specific antibodies against gangliosides[5]. The most common subtypes of GBS are Acute Inflammatory Demyelinating polyneuro-pathy (AIDP) and Acute Motor Axonal Neuropathy (AMAN), Acute Motor Sensory axonal neuropathy (AMSAN). A less common subtype is Miller Fisher Syndrome (MFS) which is characterized by ophthalmoplegia, areflexia and ataxia. Almost one third of hospitalized patientsmay progress to respiratory paralysis requiring mechanical ventilation[1].

MATERIAL AND METHODS

An observational, cross-sectional study was conducted at Government Medical College and Hospital, Aurangabad after obtaining approval from Institutional Ethics committee and valid informed consent from the patients. 41 patients who satisfied the inclusion and exclusion criteria were studied from October 2019 to October 2021. Following admission in wards, the patients were initially stabilised and after obtaining written informed consent from patient or relatives, (in cases where patient was not in a condition to consent), the patients were enrolled in the study. Detailed history was obtained regarding the onset, duration, progression and nature of symptoms, followed by detail physical examination. Patients were then assessed based on Hughes score at admission.

Table 1: Hughes Score[6]

Hughes score	0	1	2	3	4	5	6
Clinical performance	Normal	Slight clinical symptoms and signs	Able to walk 5 meters or more without assistance but unableto run	Able to walk 5 m with help	Bedridden or chair-bound	Ventilator-assisted breathing	Death

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Patients underwent CSF analysis and electrodiagnostic testing. The details were entered in the preformulated proformas. The findingswere periodically entered in the excel sheets. Based on history, examination findings and investigations patients were classified into following GBS Types:

1. Acute Inflammatory Demyelinating Polyneuropathy (AIDP)-Most common form of GBS characterized by progressive, symmetric weakness of both legs and arms, areflexia, with mild sensory signs and symptoms with cranial nerve involvement (bifacial palsies) associated with autonomic dysfunction and electrodiagnostic studies showing nerve conduction slowing or block with elevated CSF proteins.[7]
2. Acute motor axonal neuropathy (AMAN) – Pure motor axonal form of GBS.[7]
3. Acute motor and sensory axonal Neuropathy (AMSAN)-The axonal variant of GBS having both motorand sensoryinvolvement.[7]
4. Miller Fischer Variant - A variant of GBS characterized by Ophthalmoplegia, ataxia and areflexia[7]

Patients' disability scoring was done based on Hughes scale again at outcome. Patients were treated with IVIg or Plasmapheresis. Those who couldnot be given either of these treatment modalities due to financial constraints, were given supportive treatment.

Statistical Analysis

The Data was collected regularly using preformulated proformas and entered into Microsoft excel data sheet and was analyzed using SPSS 22 version software. Categorical data was represented in the form of Frequencies and proportions. Chi-square test was used as test of significance for qualitative data.

Continuous data was represented as mean and standard deviation. Wilcoxon Signed rank test is the test of significance for paired data suchas before and after for qualitative data. Kruskal Wallis test was the test of significance to identify the mean difference between more than two groups for qualitative data. Graphical representation of data: MS Excel and MS word was used to obtain various types of graphs such as bar diagram, Pie diagram.

p-value (Probability that the result is true) of <0.05 was considered asstatistically significant after assuming all the rules of statistical tests.

Statistical software: MS Excel**,** SPSS version 22 **(**IBM SPSSStatistics, Somers NY, USA) was used to analyze data.

RESULTS

Table 2: Age and sex distribution of GBS cases

Age in years	Male		Female		Total	
	Cases	%	Cases	%	Cases	%
<20	10	24.39	5	12.19	15	36.58
21 to 30	1	2.44	3	7.43	4	9.75
31 to 40	4	9.75	3	7.32	7	17.07
41 to 50	3	7.43	1	2.44	4	9.75
51 to 60	4	9.75	3	7.43	7	17.07
>60	2	4.87	2	4.87	4	9.75
Total	24	58.53	17	41.46	41	100

Mean age of subjects was 36.63 ± 19.2 years.

In this study of 41 cases, 58.53% were male and 41.46% were female. 36.58% cases belonged to younger age group of <20 years. However we have included patients with age >12 years only. (Patients with Age <12 years get admitted to the Paediatric Department) Age groups 21-30, 41-50, and > 60 years had a total of 4 cases each.

Distribution of cases were studied throughout the year. Maximum number of cases were found in the month of February(19.5%) while the least number of cases were found in the months of January and March, 2.43% each. Earlier the cases used to cluster in the monsoon season. But with climate changes seen in the past few years, this observation could not be confirmed in the present study.

Table 3: Distribution of cases according to their clinical features:Motor pattern

Motor Involvement Pattern	Cases	Percentage
Only Quadriparesis	12	29.26
Only paraparesis	10	24.39
Quadriparesis+ Facial Palsy	1	2.4
Paraparesis + Facial Palsy	2	4.87
Quadriparesis + Facial Palsy+Respiratory failure	4	9.75
Paraparesis + Facial Palsy+Respiratory Failure	0	0
Quadriparesis + Respiratory Failure	12	29.26
Paraparesis+ Respiratory Failure	0	0

At presentation, overall 26 cases (63.4%) had quadriparesis and 15 (34.1%) patients had paraparesis. Ascending paralysis was the most common presentation. Of these, only quadriparesis was seen in 12 cases (29.26%) while quadriparesis was associated with facial palsy in 1 patient (2.4%). Quadriparesis with facial palsy and respiratory insufficiency was seen in 4 cases (9.75%). 9 patients having quadriparesis developed respiratory failure while 3 cases with parapresis developed respiratory failure. Thus, overall 16 patients (39.02%) had respiratory failure. Areflexia was found in 80.48% cases, however, reflexes were present in 19.4% cases. Autonomic involvement in the form of postural hypotension was seen in 1 patient. Sensory loss was seen in 4 patients (9.8%). 1 patient in the study showed signs of cerebellar involvement had Miller Fischer variant of GBS.

Table 4: Variants of GBS

Type of GBS	Male		Female		Total	
	Cases	%	Cases	%	Cases	%
AIDP	12	29.26	13	31.7	25	60.97
AMAN	8	19.5	1	2.43	9	21.95
AMSAN	3	7.31	3	7.31	6	14.63
Miller Fischer variant	1	2.4	0	0	1	2.43

In the study 25 cases (60.97%) had AIDP. 9 cases (21.9%)were found to have AMAN variant while 6 (14.63%) cases had AMSAN variant. Only 1 case (2.43%) was found to have Miller Fischer variant of GBS.

Table 5: Functional outcome as assessed by Hughes score

Type of GBS	On admission			On discharge			P value
	Mean	SD	Median	Mean	SD	Median	
AIDP	3.68	1.14	4	2.72	1.1	3	<0.001*
AMAN	3.78	1.2	4	3.56	1.67	4	0.594
AMSAN	3.17	1.33	3	2.83	1.6	3	0.611
Miller Fischer	1.00	-	1	3.00	-	3	-

variant		
P value#	0.136	0.450

The incidence of AIDP among males and females was found to be almost similar, however AMAN was found to be more common among males (19.5%) than in females (2.43%). But the incidence of AMSAN between males and females was same, i.e. 7.3% each.

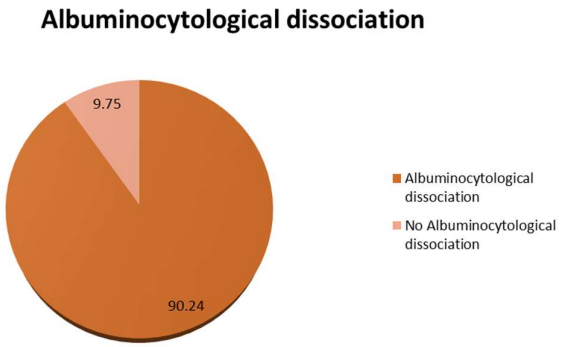

Fig 1: Pie diagram showing CSF findings in GBS cases

Albumino-cytological dissociation in CSF findings was present in 37 patients (90.24%) (Fig.1)

In the study median Hughes score at admission among subjects with AIDP was 4, among subjects with AMAN was it was 4, and among subjects with AMSAN it was 3 and among Miller Fischer variant it was 1. At discharge, median score among subjects with AIDP, AMAN, AMSAN and Miller Fischer variant was 3, 4, 3 and 3 respectively. There was statistically significant decrease in median Hughes score at discharge compared to admission in subjects with AIDP as suggested bya p-value <0.001. There was no significant difference in median Hughes score at admission and at discharge in subjects with AMAN and AMSAN.

Table 6: Outcome of patients

Treatment given	Discharge	Deaths	Total
Full dose of IVIg	15 (36.58%)	1 (2.43%)	16 (39.02%)
Incompletedose of IVIg	17 (41.46%)	3 (7.31%)	20 (48.77%)
Plasma-pheresis	1 (2.43%)	0	1 (2.43%)
Did not receiveIVIg/Plasmapheresis	2	2	4

Out of total 41 cases, 36 cases (87.8%) cases received IVImmunoglobulin either complete or a partial dose. Of these, about 16 (39.02%) cases received a fullose of IVIg, remaining 20 cases (48.78%) did not receive the complete dose of IVIg due to various constraints like lack of necessary documentation to register them under Government Scheme. Being non affording population, their doses of IVIg could not be completed. Of those patients who received IVIg, 4 patients died. Only 1 patient in the study underwent plasmapheresis who eventually improved and was discharged. 4 (9.75%) patients in the study did not receive IVIg/ Plasmapheresis due to financial constraints. 2 (2.87%) of them showed an improvement while 2 succumbed.

Table 7: Outcome of patients requiring ventilatory support:

	Discharged	Died	Total
No. of patients requiringVentilatory Assistance	10	6	16
Percentage	62.5%	37.5%	100%

In the study 16 subjects required Ventilatory assistance. Among them 62.5% were discharged and 37.5% died. Among the total deaths, 1 death was attributed to the complication of prolonged ventilation i.e. Ventilator associated Pneumonia and Septicemia while the remaining deaths were due to severe respiratory failure.

DISCUSSION

Total 41 patients of GBS were studied from August 2019 to Nov 2021 at tertiary care hospital. The mean age of patients in present study was 36.63 ± 19.2 years. We found a male preponderance (58.53%)

among the GBS cases. 36.58% cases belonged to younger age group of <20 years. However we have included patients with age >12 years only. (Patients with Age <12 years get admitted to the Paediatric Department).

Table 8: Comparison of demographics between various studies

	Present Study	Manisha Shrivastava et al[1]	Bhargavi Thota et al[16]	Sandip Sen et al[39]	Saroj Kumar Bhagat et al[2]
Year	2019-2021	2002-2013	2017-2018	2015-2018	2013-2017
Place	Maha-rashtra	Bhopal Memorial Hosp.& Res. Centre, Bhopal	Shri Venkate-shwara Institute of Medi.Sciences, Tirupati	Dr B.C.Roy, PG Institute of Pediatric Science, Kolkata	B.P. Koiral institute of Health Sciences, Nepal
No.of cases studied	41	66	64	108	31
Age group	>12 years	10 years and above		1.2 to 10 years	
Males	58.53% (n=24)	71.21%(n=47)	57.8% (n=37)	61.1% (n=66)	48.4% (n=15)
Females	41.46% (n=17)	28.78% (n=19)	42.18% (n=27)	38.8%(n=42)	51.6%(n= 16)
Mean age	36.63± 19.2 years	40.69± 18.8 years	45.9± 15.9 years	4.2 years	17 years

Table 9: Comparison of clinical features of GBS in various studies

Parameters	Present Study	Manisha Shrivastava et al[1]	Bhargavi Thota et al[16]	Sandip Sen et al[39]	Saroj Kumar Bhagat et al[2]
Quadriparesis	63.4%	74.2%	93.8%	8.3%	
Paraparesis	36.58%	25.8%	3.12%	27.8%	93.5%
Resp.Failure	36.6%	12.1%	37.5%	22.2%	16.1%
Auto.dysfunction	9.75%	0	-	-	12.9%
Areflexia	80.48%	100%	100%	94.4%	
Sens.involvement	17.1%	3%	6.25%	61.1%	22.6%

Our study results correlated with the findings in the studies carried out by Manisha Shrivastava and Bhargavi Thota. However the difference in the mean age of our study population and the study by Sandip Sen and othersat Kolkata, is attributable to the different age groups studied.

Table 10: Comparison between types of GBS in various studies

Parameters	Present Study	Manisha Shrivastava et al[1]	Bhargavi Thota et al[16]	Sandip Sen et al[39]	Saroj Kumar Bhagat et al[2]
AIDP	60.95%	50%	10.9%	52.8%	19.4%
AMAN	21.95%	43.9%	34.3%	31.48%	
AMSAN	14.63%	3%	53.1%	1.85%	19.4%
Miller Fischer variant	2.4%	0	3.1%	0	-

Diagnosis based on NCV findings- In our study the majority of cases (60.97%) were found to have AIDP variant while 22% had AMAN and 14.7% had AMSAN variant. Only 1 case (2.4%) was found to have Miller Fischer variant of GBS. The incidence of AIDP among males and females was found to be almost similar. However AMAN was found to be more common among males (19.5%) than in females (2.43%). But the incidence of AMSAN between males and females was same, i.e. 7.3% each. Only 1 male patient of Miller Fischer variant was found in the study. Thus, the findings in our study are consistent with the findings in the studies carried out by Manisha Shrivastava and Sandip Sen.

Out of total 41 cases, majority of the cases, i.e., 36 cases (87.8%) cases received IV Immunoglobulin. Of these, about 16 (39.02%) cases received a full dose of IVIg that is recommended according to weight. Remaining 20 cases (48.78%) did not receive the complete dose of IVIg. Of those patients who received IVIg, 4 patients died. Only 1 patient in the study underwent plasmapheresis who eventually improved and was discharged. 4 (9.75%) patients in the study did not receive IVIg/ Plasmapheresis and 2 (2.87%) of them showed an improvement while 2 succumbed.

In our study complete dose of IVIg could not be given to all the patients due to lack of documentation

to register them under MJPJAY (a Government scheme) and financial constraints to purchase the drug as the patients catered by this tertiary care centre, mostly belong to the lower socio economic strata. However, there was 1 death among the patients who received complete dose of IVIg owing to the complications of prolonged ventilation i.e. Ventilator associated Pneumonia and septicemia. Rest all patients who received IVIg in complete doses improved clinically.

In the study conducted by Manisha Shrivastava and others[1], all the patients underwent Plasmapheresis and all of them demonstrated clinical improvement.

Outcome - Out of 41 patients in our study, 16 (39%) patients required ventilator assistance. These patients had presented with a severe disease. 10 (24.4%) among them demonstrated recovery and were discharged while 6 (14.7%) of them expired. Of the 6 patients who succumbed, 1 death was due to the complications of Ventilator associated Pneumonia and rest of the patients had severe respiratory failure.

In the study carried out by Manisha Shrivastava and others[1], thepercentage of patients needing ventilatory assistance was as low as 1.5%. Their study subjects showed very good recovery rate with 100% discharges and no deaths. All of their patients in the study were treated with Plasmapheresis.

The percentage of patients requiring ventilatory assistance was about 23.5% and 22.2% in the studies carried out by Bhargavi Thota et al[8] and Sandip Sen et al[10] respectively. It was very close to the figures in our study population as the centers in the previous observational studies were tertiary care centers similar our study centre. However, about 91.7% patients were discharged and only 8.3% patients died in the course of study conducted by Sandip Sen and others[10]. This was similar to findings in the study carried out by Saroj Kumar Bhagat and others[9] where there were about 90% discharges with a mortality of 6.5%. The death percentage of GBS patients at our centre was relatively more than that observed in other studies. This may be due to referral bias. It can be attributed to the fact that the patients were referred late and the financial constraints for administering the standard care of treatment i.e. IVIg and plasma filter are costly and not freely available.

Summary

The present study was done at GMCH, Aurangabad in Maharashtra. It was undertaken to study clinical profile, electrophysiological profile and functional outcome of GBS patients. The study comprised of 41 cases who were diagnosed to have Guillian Barre syndrome who were >12 years of age and fulfilled the inclusion and the exclusion criteria.

- There was a slight male preponderance in our study.
- Mean age of the population under study was 36.63 ± 19.2 years. 36.58% were <20 years.
- Majority of the cases (39.02%) belonged to the lower socioeconomic class.
- Most common presentation was quadriparesis followed by paraparesis. About 39.02% of the cases had respiratory failure. Areflexia was seen in 80.48% cases. Autonomic involvement was seen in 9.8% of the cases only. Only 1 case was diagnosed to have the Miller Fischer variant of the disease.
- Of the total cases, 60.97% were diagnosed to have AIDP followed by AMAN (21.95%).
- Out of all the cases in the study, 87.8% cases received IV Immunoglobulin and 2.43% received plasmapheresis.
- 85.4% cases were discharged following recovery. 62.5% patients who had severe respiratory distress on presentation were discharged after recovery.
- There was statistically significant decrease in Hughes score at Discharge compared to admission in the patients who had AIDP as suggested by a p value <0.05.
- No seasonal variation was found in the occurrence of GBS cases.

CONCLUSION

Quadriparesis was the most common presentation among Guillian Barre syndrome patients. The majority of the patients had AIDP (Acute Inflammatory Demyelinating Polyneuropathy) subtype of GBS. There was statistically significant decrease in Hughes score in AIDP patients at discharge as compared to the Hughes score at admission after standard treatment. There was no seasonal variation in the occurrence of GBS.

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