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EXPLORING THE RELATIONSHIP BETWEEN EGRIS AND THE NEED FOR MECHANICAL VENTILATION IN GUILLAIN BARRÉ SYNDROME

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ABSTRACT

Background and objective:
Guillain-Barré syndrome (GBS) is an immune system-mediated polyradiculoneuropathy that accounts for approximately 100,000 new patients per year globally. GBS has also been linked to respiratory failure in 20% to 40% of cases. The objective of this study was to find an association between Erasmus GBS respiratory insufficiency score (EGRIS) and the need for mechanical ventilation.

Methods:
We conducted our cross-sectional study at department of Neurology, Pakistan Institute of Medical Sciences from November 2019 to February 2020. Sixty patients were selected for this study. Data including demographic profile, variant of GBS, and EGRIS was calculated from all the study participants. Data was entered and analyzed using SPSS version 25.

Results:
All 60 patients enrolled in the study underwent continuous monitoring during admission time. Out of 60 patients, three (5%) endured mechanical ventilation with a mean EGRIS of 4.5. Others who didn't need mechanical ventilation had mean EGRIS of 1.5. The three patients requiring mechanical ventilation were one each from Acute Inflammatory Demyelinating polyradiculoneuropathy, Acute Motor Axonal Neuropathy, and Acute Motor and Sensory Axonal Neuropathy variants of GBS. p-value was non-significant for the presented data.

Conclusion:
EGRIS could not meet statistical significance in identifying the requirement of mechanical ventilation for GBS in this study. This may be due to low sample size.

Keywords: EGRIS, Mechanical ventilation, GBS, Neuropathy

INTRODUCTION

Guillain-Barré syndrome (GBS) is an immune system-mediated polyradiculoneuropathy that accounts for approximately 100,000 new patients per year globally.[1] GBS is also linked to respiratory failure in 20% to 40% of cases.[2] To predict respiratory muscle involvement is vital to avoid respiratory distress and prioritize cases in the intensive care unit (ICU). Optimum management is required in GBS to avoid respiratory failure so prediction of its clinical features and in-time intubation of a patient is required. Swift intervention is linked with decreased rates of complications and improved results for the patient.[3] In most cases, acute onset of neurological symptoms follows an infective disease subsequently causing advanced limb weakness that could progress for four months before hitting a plateau. Various infections are linked to GBS. Among these Campylobacter jejuni is found to be the most common and is reported by various researchers. For C jejuni associated GBS, there are strong pieces of evidence that suggest that molecular mimicry is present among nerve endings and

microbial antigens, causing the development of GBS.[5] The classic manifestation of GBS does not generally cause a diagnostic challenge, but atypical forms are overlooked when not examined for. For assistance in the diagnosis, nerve conduction studies are carried out to detect polyradiculoneuropathy and cerebrospinal fluid examination could reveal albumin cytological dissociation. Yet diagnosis should never solely depend on these tests because both of these could be normal in the initial stage of the syndrome.[6] Patients suffering from GBS need rigorous monitoring for disease advancement, specifically for autonomic dysfunction, respiratory insufficiency, and bulbar weakness. Predictive scales for the prediction of patient outcomes and to classify treatment have been developed. Plasma exchange and intravenous immunoglobulin are the only two approved immunotherapies that can speed up recovery in GBS. Even after treatment via standard immune therapies, approximately 5% of patients die, while approximately 20% of patients cannot not walk unassisted at least for 12 months after the disease progression.[6]

In studies done by various Intergovernmental Organizations (IGOs), an antecedent incident was identified in 76% of cases, primarily nose, sinuses, mouth, pharynx, and larynx infections (35%) in East Asia and Southeast Asia, North America, and Europe, while intestinal infection (27%) was more prevalent in Bangladesh.[7] GBS has also been linked to certain vaccination sand with immune checkpoint inhibitor therapy.[8,9] Moreover less frequent triggers consist of ganglioside application and surgery. In 1991, McKhann and colleagues researched the seasonal epidemic of acute equivocal Chinese paralysis syndrome.[10] This hallmark research afterward led to the identification of acute motor axonal neuropathy (AMAN) and acute motor-sensory axonal neuropathy (AMSAN) as clinical variants of GBS caused by C jejuni infection. Up till now, different types of GBS variants had been identified. Most prevalent of these are AMSAN and AMAN (axonal types) and acute inflammatory demyelinating polyneuropathy (AIDP).

Erasmus GBS Respiratory Insufficiency Score (EGRIS) gave 91% predictive value for mechanical ventilation (MV) at the time of admission for seven patients in one study.[11] Present study was conducted to find an association of EGRIS with the requirement of mechanical ventilation (MV) and to verify its effectiveness as a predictive model.

METHODS

Study design: Cross-sectional study.

Place and duration of study: This study was conducted for four months (from November 2019 to February 2020) in the Department of Neurology, Pakistan Institute of Medical sciences.

Sample size: Sixty, calculated using WHO sample size calculator.

Sampling technique: Non-probability consecutive sampling.

Inclusion Criteria: Patients aged above 16 years with acute onset ascending paralysis with NCS evidence of acute neuropathy and/or CSF analysis showing albuminocytologic dissociation.

Exclusion criteria:

- Patients with other systemic diseases that may cause neuropathy such as uremia, malignancy.
- Patients with pyramidal signs such as up-going plantar reflexes.
- Patients with clinical/radiological evidence of myelopathy

Data collection: After taking informed consent. all the 60 patients' data was collected including demographics, clinical diagnosis and GBS variant. and need for mechanical ventilation. EGRIS was calculated on all patients upon admission to Department of Neurology. The EGRIS measures the chances of MV in GBS during the first week of monitoring. It is calculated on 3 major criteria. Each criterion is considered and allotted a separate score; the addition of these scores provides total EGRIS for a patient (between 0 and 7). A score of 0–2 Predicts a low chance of mechanical ventilation (MV), 3–4 shows a moderate chance for MV and ≥5 depicts an elevated chance for MV.

Data analysis: The data was analyzed through SPSS 22.0.

Ethical considerations: The research protocol was approved by the PIMS Ethics Review committee.

RESULTS

All patients were admitted to neurology ward and underwent monitoring for clinical deterioration and respiratory distress. One predictive factor of a patient's need for early MV was the number of days between the onset of motor symptoms and hospitalization. Out of 60 patients, three (5%) went on mechanical ventilation with a mean EGRIS of 4.5. While others who didn't need mechanical ventilation had EGRIS mean of 1.5. The three patients requiring MV were one from each from AIDP, AMAN, and AMSAN. In our study 31 patients were male and 29 were female Figure 1 is a pie chart showing the age distribution in our patients with the highest number found in age group 36-45 years.

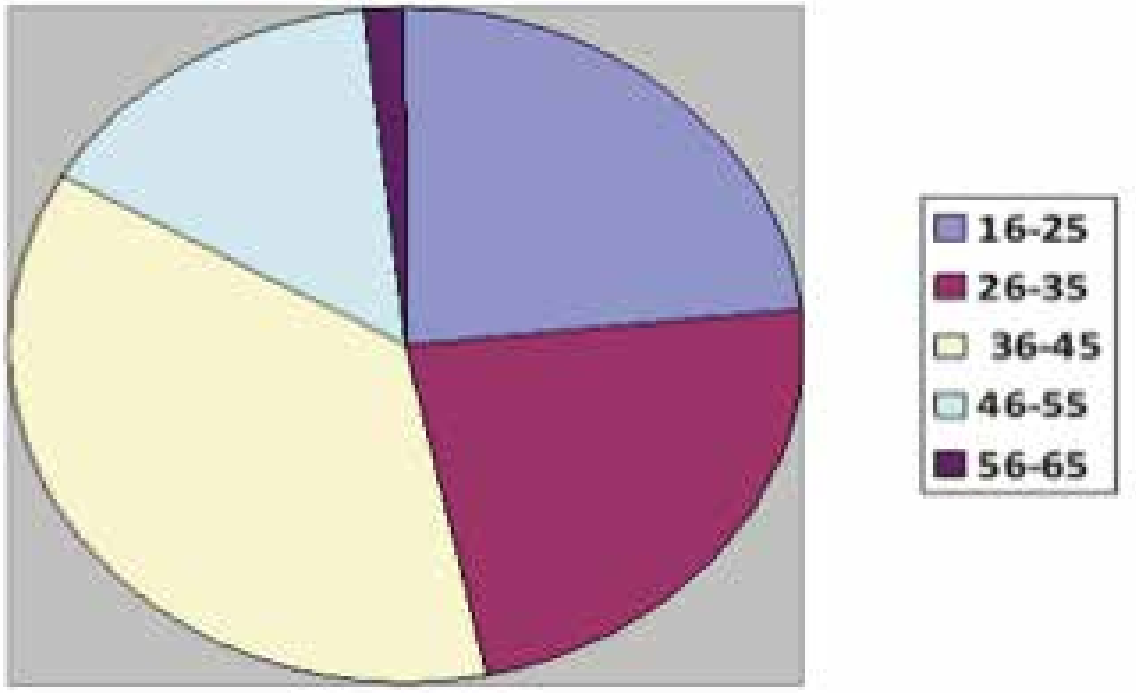

Figure 1: Illustration of age-wise frequency of patients

The highest number of patients were suffering from AMAN. The number of patients suffering from AMAN was 33 (54.1%), the number of patients suffering from ADP was 15 (24.6%) and the number of patients suffering from AMSAN was 12 (19.7%). The number of patients suffering from seven days or more was 36 (59.0%), the number of patients suffering from four to seven days was 20 (32.8%) and the number of patients suffering from less than three days was 4 (6.6%). When EGRIS was calculated for patients included in the study, 75.4% of cases had a score ≥3. Each of the three variants had the same 1.66% for the need of MV with a cumulative percentage of 5%. Details of the frequency of MV requirements in different GBS variants are given in Table 1.

Table. 1: Details of frequency of MV requirement in different GBS variants

			Need for Mechanical	ventilation	
			No	Yes	Total
GBS Variant	AIDP	Count	14	1	15
		% Need for/ No need of Mechanical ventilation	23.33%	1.66%	25.0%
	AMAN	Count	32	1	33
		% Need for/ No need of Mechanical ventilation	53.33%	1.66%	55.0%
	AMSAN	Count	11	1	12
		% Need for/ No need of Mechanical ventilation	18.33%	1.66%	20.0%
Total		Count	57	3	60
		% Need for/ No need of Mechanical ventilation	95.0%	5.0%	100.0%

AIDP: Acute inflammatory demyelinating polyradiculoneuropathy
AMAN: Acute motor axonal neuropathy
AMSAN: Acute motor and sensory axonal neuropathy

Statistical analysis of data didn't show any significant relationship between EGRIS and the need for MV (Table 2)

Table. 2: Details of statistics applied to obtained data

	Value	Degree of freedom	Asymptotic significance (2-sided)
Pearson Chi-Square	0.638	2	.727
Likelihood Ratio	.627	2	.731
Linear-by-Linear Association	.017	1	.895
N of Valid Cases	60		

DISCUSSION

GBS is a severe, immune system-mediated attack on the peripheral nervous system which results in progressive motor weakness and/or sensory loss. Its incidence rate is 0.4–4.0 per 100,000 people/year and after the decrease in the rate of polio cases now it is the largest reason for acute neuromuscular paralysis.[12] On the basis of different characteristics like clinical, pathological, and electrophysiological it is declared as a heterogeneous disorder that includes different subtypes: AIDP, AMAN, AMSAN, and Miller-Fischer syndrome.[13] EGRIS is a predictive model to estimate the early requirement of MV.[14] GBS has many electrophysiological subtypes and predominant phenotypes in various regions so the disease severity and the EGRIS also changes with change in the region.[15]

The majority of patients included in this study were male belonging to an age group of 36 to 45 years. Globally, researches from Latin American, and European literature reports its prevalence majorly in

male and aged patients.[15] In populations like Asian, and Latin American lineage AMAN was reported as a prevalent variant.[15,16] In the present study AMAN was more prevalent than AMSAN and AIDP which makes it also a prevalent variant in the Pakistani population. Of the cases in the present study, only 5% of patients required MV, which is a much lower percentage as compared to 20–25% reported globally.[16]

Two major causes of mortality in GBS are respiratory failure and autonomic dysfunction.[17,18] Treatment for GBS aims to reduce mortality and hasten the recovery from acute neuropathy that lead to paralysis. The two main treatment options apart from supportive care are therapeutic plasma-pharesis and intravenous immune globulins.[19] These treatments aim to reduce the respiratory muscle involvement and improve the survival. There are various clinical and neuro-physiological factors whose presence warns of the development of respiratory weakness.[20] For this purpose various prediction scores have been established, one of them is EGRIS. It aims to predict the development of respiratory insufficiency in GBS patients.

In the present study, it was noticed that 74.5% of patients' EGRIS was less than or equal to 3. Hence, they did not require MV. Only 3 patients whose EGRIS was greater than or equal to 5 required MV. Our results correlate with another research where higher EGRIS was associated with the early requirement of MV. Researchers from Japan and Malaysia reported the same evidence by depicting statistically significant differences in EGRIS in patients who required and did not require MV. Correspondingly, a team of researchers in South Korea reported a positive correlation between the chances of MV and EGRIS.[20]

Limitations: This study showed higher EGRIS for patients needing mechanical ventilation but due to smaller sample size it didn't reach any statistical significance.

CONCLUSION

EGRIS could not significantly predict the need for MV in GBS patients of this study. This was likely due to the small sample size. More studies are needed with larger samples to correctly analyze the utility of EGRIS.

REFERENCES

1. Sejvar JJ, Baughman AL, Wise M, Morgan OW. Population incidence of Guillain-Barré syndrome: a systematic review and meta-analysis. Neuroepi demiology. 2011;36(2):123-133. doi:10.1159/000324710
2. Lawn ND, Fletcher DD, Henderson RD, Wolter TD, Wijdicks EF. Anticipating mechanical ventila tion in Guillain-Barré syndrome. Arch Neurol. 2001;58(6):893-898. doi:10.1001/arch neur.58.6.893
3. Wijdicks EF, Borel CO. Respiratory management in acute neurologic illness. Neurology. 1998;50(1):11-20. doi:10.1212/wnl.50.1.11
4. Wachira VK, Peixoto HM, de Oliveira MRF. System atic review of factors associated with the development of Guillain-Barré syndrome 2007-2017: what has changed? Trop Med Int Health. 2019;24(2):132-142. doi:10.1111/t mi.13181
5. Yuki N, Susuki K, Koga M, Nishimoto Y, Odaka M, Hirata K, et al. Carbohydrate mimicry between human ganglioside GM1 and Campylobacter jejuni lipooligosaccharide causes Guillain-Barre syndrome. Proc Natl Acad Sci U S A. 2004;101(31):11404-11409. doi:10.1073/pn as.0402391101
6. Willison HJ, Jacobs BC, van Doorn PA. Guillain-Barré syndrome. Lancet. 2016;388(10045):717-727. doi:10.1016/S0140-6736(16)00339-1
7. Doets AY, Verboon C, van den Berg B, Harbo T, Cornblath DR, Willison HJ, et al. Regional variation of Guillain-Barré syndrome. Brain. 2018;141(10):2866-2877. doi:10.1093/brain/awy232
8. Schonberger LB, Bregman DJ, Sullivan-Bolyai JZ, keenlyside RA, Ziegler DW, Retailliau HF, et al. Guillain-Barre syndrome following vaccination in the National Influenza Immunization Program, United States, 1976--1977. Am J Epidemiol. 1979;110(2):105-123. doi:10.1093/oxfordjour nals.aje.a112795
9. Kao JC, Liao B, Markovic SN, Klein CJ, Naddaf E, Staff NP, et al. Neurological Complications Associated With Anti-Programmed Death 1 (PD-1) Antibodies [published correction appears in JAMA Neurol. 2017 Oct 1;74(10):1271]. JAMA Neurol. 2017;74(10):1216-1222. doi:10.1001/jamaneurol.2017.1912

10. McKhann GM, Cornblath DR, Ho T, Li CY, Bai AY, Wu HS, et al. Clinical and electrophysiological aspects of acute paralytic disease of children and young adults in northern China. Lancet. 1991;338(8767):593-597. doi:10.1016/0140-6736(91)90606-p
11. Walgaard C, Lingsma HF, Ruts L, Drenthen J, van Koningsveld R, Garssen MJP, et al. Prediction of respiratory insufficiency in Guillain-Barré syndrome. Ann Neurol. 2010;67(6):781-787. doi:10.1002/ana.21976
12. Bragazzi NL, Kolahi AA, Nejadghaderi SA, Lochner P, Brigo F, Naldi A, et al. Global, regional, and national burden of Guillain-Barré syndrome and its underlying causes from 1990 to 2019. J Neuroinflammation. 2021;18(1):264.
13. Dimachkie MM, Barohn RJ. Guillain-Barré syndrome and variants. Neurol Clin. 2013;31(2):491-510. doi:10.1016/j.ncl.2013.01.005
14. Kumar M, Kalita J, Kant Misra U, Dhar N. Predic tion models for mechanical ventilation and outcome in Guillain-Barré syndrome. J Clin Neurosci. 2021;92:131-135. doi:10.1016/j.jocn.2021.07.060
15. Malaga M, Rodriguez-Calienes A, Marquez-Naka matsu A, Recuay K, Merzthal L, Bustamante-Pay tan D, et al. Predicting Mechanical Ventilation Using the EGRIS in Guillain-Barré Syndrome in a Latin American Country [published correction appears in Neurocrit Care. 2021 Oct;35(2):603]. Neurocrit Care. 2021;35(3):775-782. doi:10.1007/s12028-021-01218-z
16. Zheng P, Tian DC, Xiu Y, Wang Y, Shi FD. Incidence of Guillain-Barré syndrome (GBS) in China: A national population-based study. Lancet Reg Health West Pac. 2022;18:100302.
17. van den Berg B, Bunschoten C, van Doorn PA, Jacobs BC. Mortality in Guillain-Barre syndrome. Neurology. 2013;80(18):1650-1654. doi:10.1212/WNL.0b013e3182904fcc
18. Zaeem Z, Siddiqi ZA, Zochodne DW. Autonomic involvement in Guillain-Barré syndrome: an update. Clin Auton Res. 2019;29(3):289-299. doi:10.1007/s10286-018-0542-y
19. Liu S, Dong C, Ubogu EE. Immunotherapy of Guillain-Barré syndrome. Hum Vaccin Immunother. 2018;14(11):2568-2579. doi:10.1080/21645515.2018.1493415
20. Durand MC, Porcher R, Orlikowski D, Aboab J, Devaux C, Clair B, et al. Clinical and electrophysiological predictors of respiratory failure in Guillain-Barré syndrome: a prospective study. Lancet Neurol. 2006;5(12):1021-1028. doi:10.1016/S1474-4422(06)70603-2

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Anum Irfan; Data collection, data analysis, manuscript writing
Waleed Malik; Data collection, data analysis, manuscript writing
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All the authors have approved the final version of the article, and agree to be accountable for all aspects of the work.