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Correlation of Modified EGRIS with the Risk of Mechanical Ventilation in GBS

Dr. Sohini Ganji[1], Dr. Veena Narisetty[2], Dr. Akhilesh Kumar[3], Dr. A. Goutami Priyadarshini[4]

[1]Osmania Medical College
Email: *drsohiniganji[at]gmail.com*

[2]Osmania Medical College
Corresponding Author Email: *veenanarisetty[at]yahoo.com*

[3]Osmania Medical College
Email: *mailyourdoc[at]gmail.com*

[4]Osmania Medical College
Email: *docgoutami[at]gmail.com*

Abstract: ***Introduction: Guillain–Barré syndrome (GBS) is an acute onset rapidly progressive disease due to inflammation of the peripheral nervous system and is one of the commonest causes of flaccid paralysis. Involvement of the respiratory muscles causes the life-threatening complication of respiratory failure requiring the need of mechanical ventilation, seen in up to 22% within the first week. Prompt recognition of these patients at increased risk of respiratory failure in GBS is needed to triage them and shift them timely to a respiratory intensive care unit. Modified Erasmus GBS Respiratory Insuffiency Score (mEGRIS) is a bedside clinical score based on 4 parameters, duration of onset of weakness until admission, presence or absence of bulbar palsy, neck power and power of bilateral hip flexion. It can be used to predict MV risk at different time points during admission course and is applied in all GBS variants. Here we present 56 cases of GBS where the mEGRIS score was used to predict the risk of MV. Clinical Methods: 56 patients who met the criteria for GBS and admitted in Neurology Department were enrolled. They were evaluated by detailed examination, routine investigations, CSF and NCS. The mEGRIS score was used to triage patients who require MV. Results: In our study, out of 56 patients, 26 were male and 30 were female. AMSAN was MC presentation in 38 patients. 19 had bulbar involvement, 16 required MV out of which 7 patients died. The cause of death was septic shock in 2 patients, PTE in 1 patient and autonomic fluctuations in 1 patient. 2 patients succumbed to underlying comorbidities. All patients who required MV had high mEGRIS score at presentation. Conclusion: The mEGRIS is a rapid score that predicts the risk of MV in GBS which helps in triaging patients.***

Keywords: GBS, mechanical ventilation, respiratory insufficiency, bulbar weakness, triage

1. Introduction

Guillain–Barré syndrome (GBS) is an acute onset rapidly progressive disease due to inflammation of the peripheral nervous system and is one of the commonest causes of flaccid paralysis. It causes multifocal inflammatory demyelination of spinal roots and peripheral nerves.[1] The incidence is 1–2 per 100,000 years approximately.[1] Males were found to be more affected than females (1.5:1).[1] Patients usually reach maximum disability within 2 weeks. Involvement of the respiratory muscles causes the life threatening complication of respiratory failure requiring the need of mechanical ventilation, seen in up to 22% within the first week.[2] Delaying intubation in these patients can lead to aspiration pneumonia and sepsis, leading to worse outcomes.[3] A predictive model for the need of respiratory support is most useful if it is based on clinical features and if it can be assessed early at presentation. Strong predictors include features like rapid disease progression, bulbar or neck weakness and severe muscle weakness on hospital admission.[4]The original Erasmus GBS respiratory insufficiency score (EGRIS) incorporates three key factors: the duration between the onset of weakness and hospital admission, the presence or absence of facial and/or bulbar weakness and the Medical Research Council (MRC) sum score, which gauges muscle weakness severity. This scoring system ranges from 0 to 7 and estimates the risk of respiratory failure during the first week, with probabilities ranging from 1% to 90%.[5]. Identifying patients at high risk early is vital for appropriate triage and more intensive monitoring. The modified EGRIS (mEGRIS) is based on four clinical features, duration of onset of weakness until admission, presence or absence of bulbar palsy, neck power and power of bilateral hip flexion [6] Neck flexion strength was not included in the EGRIS and this modified version requires testing muscle power only in the proximal lower limbs making it an easier and rapid bed side score to calculate. It can be used to predict MV risk at different time points during admission course and is applicable in all GBS variants and even milder cases.[6] Here we present 56 cases of GBS where the mEGRIS score was used to predict the risk of mechanical ventilation.

Aim

To evaluate the effectiveness of the modified EGRIS score as a rapid, bedside, accurate predictor of the risk of mechanical ventilation in GBS.

2. Methodology

- **Study Design:** A prospective observational study done in the Department of Neurology, Osmania General Hospital, Telangana.
- **Place of Study:** Department of Neurology, Osmania General Hospital. A tertiary care centre in Hyderabad, Telangana.

- **Sample Size:** 56 patients who met the National Institute of Neurological Disease and Stroke criteria (NINDS) for GBS were enrolled in the study after taking informed consent.
  Inclusion Criteria: All patients who met the NINDS criteria for GBS were included in the study.
  Exclusion Criteria: Patients with GBS who were brought to the hospital on mechanical ventilation were excluded from the study.

Data Collection

Details of clinical and demographic data were collected. Complete neurological examination, routine blood investigations, CSF analysis and nerve conduction studies were done.

- The modified EGRIS (mEGRIS) score ranges from 0 to 32 as illustrated in table 1. The probability of a patient to be mechanically ventilated within 1day, 3days and 1week from admission was estimated based on the predictive probability curves from the study by Luijten et al [6] as seen in figure 1.
- The modified EGRIS score was calculated at admission for all patients.
- Patients with a high score and predictive value for requiring mechanical ventilation were admitted to the respiratory intensive care unit for further management.

Statistical Analysis

The data is presented in the form of percentages.

- Bar diagrams and pie charts are used for representation. P value is calculated using the T test to assess the association of mEGRIS with risk of mechanical ventilation in GBS.
- mEGRIS score.

Bulbar weakness	Yes	5
	No	0
Time from weakness to admission (days)	0	7
	1	6
	2	5
	3	4
	4	3
	5	2
	6	1
	>/=7	0
Neck flexion MRC (0-5)	0	10
	1	8
	2	6
	3	4
	4	2
	5	0
Bilateral Hip Flexion MRC (0-10)	0	10
	1	9
	2	8
	3	7
	4	6
	5	5
	6	4
	7	3
	8	2
	9	1
	10	0
Total		32

Table 1- modified eGRIS (he score has been used after taking written permissions from Mr Bart Jacobs, Luijten LW, Doets AY, Arends S, Dimachkie MM, Gorson KC, Islam B, Kolb NA, Kusunoki S, Papri N, Waheed W, Walgaard C. Modified Erasmus GBS Respiratory Insufficiency Score: a simplified clinical tool to predict the risk of mechanical ventilation in Guillain-Barré syndrome. Journal of Neurology, Neurosurgery & Psychiatry)

Predictive probabilities of MV

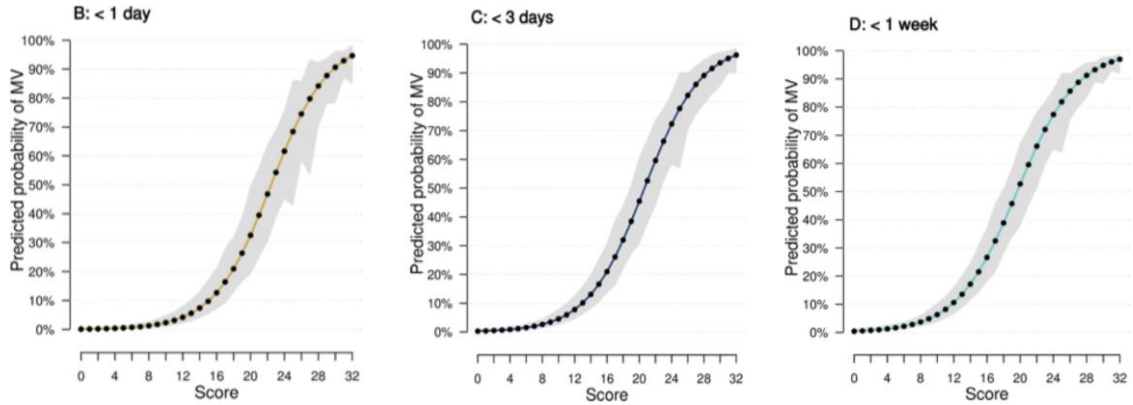

Figure 1: Predictive probability curves (the graphs have used after taking written permissions from Mr Bart Jacobs, Luijten LW, Doets AY, Arends S, Dimachkie MM, Gorson KC, Islam B, Kolb NA, Kusunoki S, Papri N, Waheed W, Walgaard C. Modified Erasmus GBS Respiratory Insufficiency Score: a simplified clinical tool to predict the risk of mechanical ventilation in Guillain-Barré syndrome. Journal of Neurology, Neurosurgery & Psychiatry)

3. Results

Sex distribution- Out of 56 patients, 26(46%) were males and 30 (54%) were females.

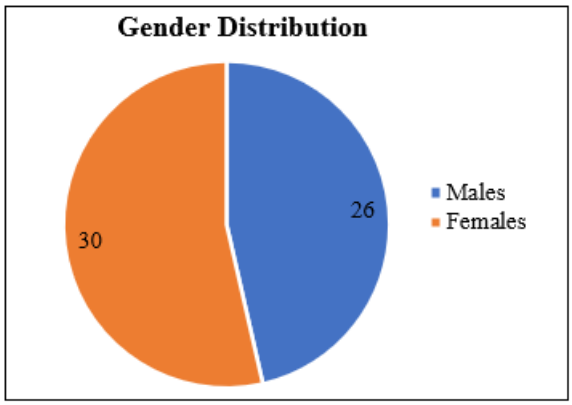

Figure 2: Sex Distribution

- Age distribution of patients- Most patients were in the > 40 sub group (48.2%). 8.9 % were less than 20 years and 42.8% were in the 20-40 years sub group.
- GBS- disability score at presentation- 21(37.5%) patients had Hughes grade 4 and 14 patients had Hughes grade 5B.

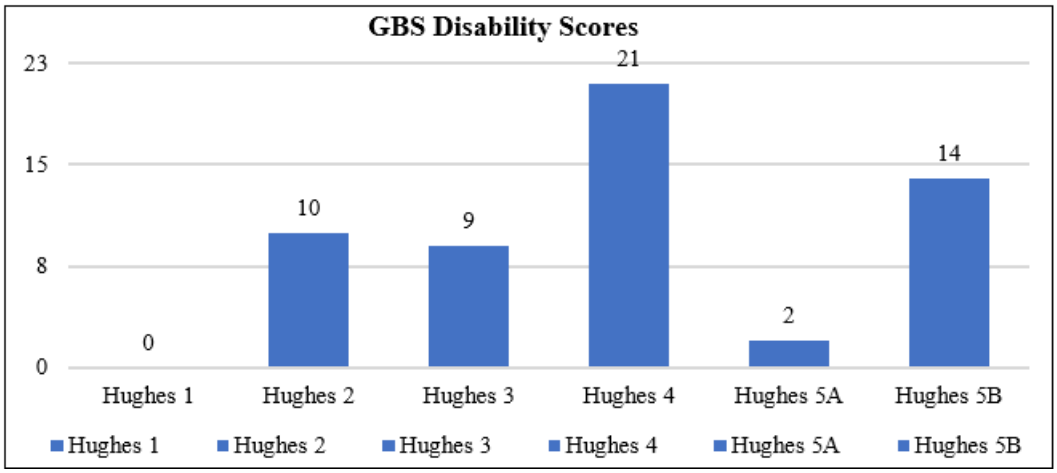

Figure 3: GBS- disability score at presentation

- GBS subtypes- 38 patients had AMSAN variant of GBS and was the most common subtype in our series. 2 patients had atypical presentation and was diagnosed as having a Miller- Fischer variant.

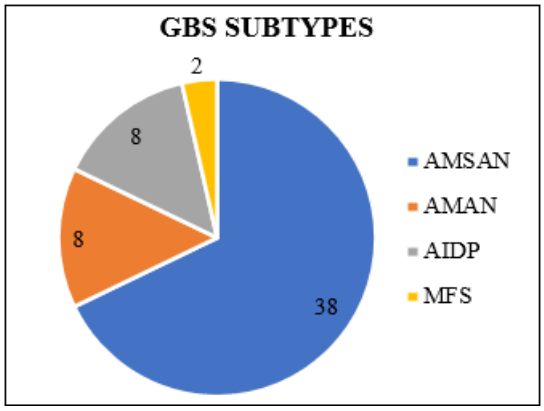

Figure 4: GBS subtypes

- Two females were pregnant, one of them presented at 20 wks of gestation and had a milder phenotype needing only conservative management. The other patient presented at 36 wks with Hughes grade 4, without requiring mechanical ventilation and was treated with IVIG.
- Facial palsy was present in 14 patients (25%) and autonomic fluctuations were present in 6 patients (10.7%). Neck flexion power of MRC 3 or less was seen in 15(26.7%) patients and 73% of them required MV.
- Mechanical Ventilation- 19(33.9%) patients had bulbar involvement and 16(28.5%) of them required ventilator support.
- All 40 (71.4%) patients without bulbar involvement are doing well. 7 (12.5%) patients who required mechanical ventilation expired.
- Out of the patients who expired, 2 patients died because of severe autonomic fluctuations, 2 patients had severe sepsis, one succumbed to acute pulmonary thromboembolism, one patient had underlying heart failure with reduced ejection fraction and one had chronic kidney disease requiring plasmapheresis and developed refractory shock. 2 of the patients who died were less than 20 years old.
- The details of the patients who required ventilatory support is illustrated in table 2.

Table 2: Demographics of patients requiring mechanical ventilation

Age	Sex	mEGRIS score	Risk of MV at day 1, day 3 and 1 week	Treatment	Outcome	Comorbidities	
38	M	17/32	20%, 25%, 30%	IVIG	Improved	DM, HTN	
53	F	24/32	60%, 70%, 75%	IVIG	Improved	HTN	
21	M	26/32	70%, 75%,80%	IVIG	Improved	None	b/l facial palsy
12	M	18/32	25%,30%, 35%	IVIG	Improved	None	b/l facial palsy
24	F	25/32	62%, 67%, 72%	IVIG, IVMP	Expired	None	b/l facial palsy, Acute PTE
55	M	23/32	55%, 60%,70%	IVIG	Improved	HTN, DM	B/l ptosis, ophthalmoplegia, b/l facial palsy, lung malignancy
20	F	24/32	60%, 70%, 75%	IVIG, IVMP	Expired	None	Severe autonomic fluctuations
40	F	25/32	62%, 67%, 72%	IVIG, IVMP	Improved	HTN, DM	
40	M	26/32	70%, 75%,80%	IVIG	Expired	HTN	Aspiration pneumonia, Sepsis
19	M	17/32	20%, 25%, 30%	PLEX	Expired	CKD, glomeru-lonephritis	Refractory shock
27	M	6-32	2%	PLEX	Improved	CKD on MHD, DM, HTN	
54	M	29/32	80%, 85%, 90%	IVIG	Expired	CAD s/p PTCA	Heart failure, MI
55	M	23/32	55%, 60%,70%	IVIG	Expired		Sepsis, Aspiration pneumonia
35	F	27/32	75%, 80%, 85%	IVIG, IVMP	Expired		Acute PTE, Severe autonomic fluctua-tions, b/l facial palsy
64	M	21/32	40%, 45%, 50%	IVIG, IVMP	Improved		Autonomic fluctuations, Right facial palsy

Treatment- 30(51.7%) patients were treated with IVIG, 6 (10.7%) patients received both IVIG and IVMP in view of autonomic fluctuations, 2(3.5%) patients received plasmapheresis and 18 (32%) patients were managed conservatively.

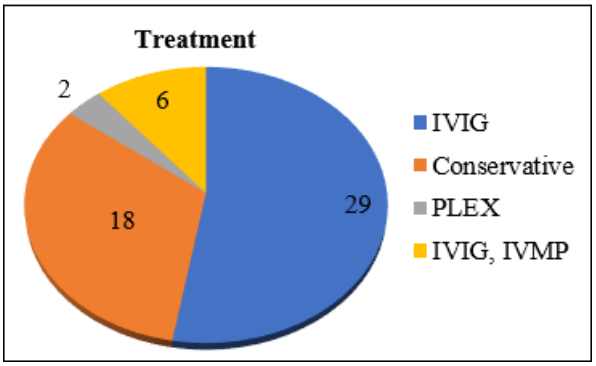

Figure 5: Treatment groups

The t test was applied to calculate the association between the mEGRIS score and risk of mechanical ventilation and the p value (0.00001) was statistically significant.

Table 3: Association between EGRIS Score with requirement of Mechanical ventilation

mEGRIS Score	Requirement of Mechanical ventilation		P value
	Yes	No	
Mean	22	6	0.00001*
Standard Deviation	5.71	4	

A higher score was also associated with a poorer outcome (p value < 0.00001).

Table 4: Association between EGRIS Score with Outcome

mEGRIS Score	Outcome		P value
	Improved	Expired	
Mean	9.0	24.0	0.0001*
Standard Deviation	6.73	3.82	

4. Discussion

- Respiratory failure develops insidiously in GBS without the usual signs of respiratory compromise.[5] Early prediction of mechanical ventilation is important as it will help in reducing the risk of complications and improving outcomes.
- In our study females were more commonly affected than males. Majority of the patients were older than 20 years and AMSAN was the most common GBS variant in our study.
- A study of demographics and clinical profile of GBS patients in central India showed a male preponderance with AIDP being the most common clinical subtype.[7]
- In our study, facial palsy was present in 25% of patients and bulbar involvement was seen in 33.9% of patients and almost all of them required MV. A study from china found shorter time from onset to admission, facial and bulbar palsy and lower MRC sum score at presentation were associated with early MV.[8]
- Electrophysiological parameters like an early conduction block greater than 50% in the distal peroneal nerve has been strongly linked to the requirement for mechanical ventilation.[6]
- A neck flexion score of 3 or less was seen in 26.7% of patients and 73% of them required intubation. This is in concordance with a study done by Lisa M Arnold et al who reviewed the relationship between neck flexion and need for intubation and found that a MRC score less than 3 was strongly associated with risk.[9]
- Gallardo E et al. reported on ultrasonographic changes in Guillain-Barré Syndrome (GBS) and observed that patients with cervical spinal root involvement had a higher likelihood of developing respiratory failure. Their findings suggest that assessing neck muscle strength might serve as an indirect indicator of the risk for respiratory failure.[10]
- In our study all the patients requiring mechanical ventilation had very low MRC sum scores at presentation and

rapid progression of disease suggesting that these had direct correlation with risk of mechanical ventilation similar to other studies.

- The EGRIS predicts the probability of needing mechanical ventilation within a week of admission. This streamlined score involves evaluating just one muscle group and includes the assessment of neck flexion.
- The score also gives predictive probabilities of mechanical ventilation at multiple time points during admission. A high score also correlated with a worse outcome.
- Thus, this score can be used as an effective bedside clinical parameter to shift patients to a respiratory intensive care unit for strict monitoring and elective intubation.

5. Limitations

The score was not externally validated.

- Small sample size.
- Needs larger studies and different clinical settings to suggest effectiveness and applicability of the score.

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