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# Guillain–Barré syndrome from an emergency department view: how to better predict the outcome?

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**ABSTRACT**

**Objective:** In Guillain–Barre syndrome (GBS), respiratory failure is the most serious manifestation and mechanical ventilation (MV) is required in approximately 20% of the patients. In this retrospective study, we aimed to evaluate clinical factors that can be evaluated in the Emergency Department which may influence the short-term prognosis of GBS patients.

**Methods:** Data were acquired regarding age, sex, antecedent infections, neurological signs and symptoms, cerebrospinal fluid examination, nerve conduction studies, treatment of GBS, need for MV, length of stay in the hospital, and discharge destination (home or rehabilitation). Charlson Comorbidity Index and modified Erasmus GBS outcome score (mEGOS) were collected on admission.

**Results:** Seventy-eight GBS patients were recruited with a mean age of 53.9 (range 19-81). Sixty-nine (88.46%) were diagnosed with GBS and nine (11.54%) had classic Miller-Fisher syndrome. Mean values for the Charlson Comorbidity index were 1.20 ± 1.81, and the values of mEGOS were 2.4 ± 1.6. The rate of home discharge and rehabilitation was similar between elderly and younger patients. Patients who required MV had higher mEGOS (p-value=0.061). Regarding the electrophysiological subtypes, we did not observe a significant difference between AIDP and AMAN/AMSAN concerning the need for MV, the type of discharge, values of mEGOS and Charlson Comorbidity Index.

**Discussion:** A significant correlation was found between mEGOS and the need for MV. Age did not influence the short-term prognosis of GBS patients. mEGOS may be a useful tool for predicting outcomes in patients with GBS and higher mEGOS scores on admission significantly correlated with poor outcomes.

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Guillain–Barré syndrome; mEGOS; Charlson comorbidity index; mechanical ventilation

## Introduction

Guillain–Barre syndrome (GBS) is a post-infectious, immune-mediated neuropathy characterized by rapidly evolving ascending weakness, mild sensory loss, and hypo- or areflexia, progressing to a nadir within 4 weeks [1].

infection

Diagnosis of GBS is based on the patient history and neurological, electrophysiological, and cerebrospinal fluid (CSF) examinations [2,3].

Treatment of GBS usually includes supportive medical care and immunotherapy such as intravenous immunoglobulin (IVIg) and plasma exchange (PE) [4].

Even with modern intensive care and IVIg treatment, GBS has an associated mortality rate between 3% and 7% in the acute phase and a residual disability rate of 20% or more. Respiratory failure is the most serious manifestation of GBS, and mechanical ventilation (MV) is required in approximately 20% of patients [5]. Early identification of GBS patients with respiratory failure in need of respiratory support is crucial as early intubation for GBS patients seems to be beneficial [6]. However, a significant proportion of patients with GBS receiving MV ultimately require tracheostomy throughout the disease course [7].

Many prognostic factors correlated with poor long-term outcomes have been proposed, such as older age of the patients, the need for MV, and lower MRC sum score on admission [8–10]

The Erasmus GBS group developed and revised a prognostic model to combine some predictive factors referred to as modified Erasmus GBS outcome score (mEGOS) to predict the functional outcome at 6 months. The mEGOS can be applied already at hospital admission, and it is calculated considering age, presence of diarrhea before the onset of symptoms, and severity of muscle weakness assessed through the Medical Research Council (MRC) sum score [11].

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The Charlson Comorbidity Index accounts for multiple comorbidities by creating a sum score weighted according to the presence of comorbid conditions that alter the risk of 1-year mortality [12,13].

Approximately, 40% of the patients with GBS who are hospitalized benefit from a multi-disciplinary rehabilitation, which is essential in the recovery [14].

In the current study, we aimed to evaluate factors that can be easily and early assessed in Emergency Department, which may influence the prognosis of GBS patients.

## Methods

### Study design

This is a monocentric, retrospective study conducted in a teaching urban hospital with an annual attendance at the ED of about 75,000 patients (more than 87% adults). After approval by our institution review board, all clinical records of consecutive patients ≥18 years admitted to the Emergency Department of Fondazione Policlinico Universitario A. Gemelli IRCCS in Rome, from 1 January 2014 to 31 December 2018, have been evaluated and patients with a discharge diagnosis of GBS in this period were recruited.

### GBS diagnosis

Diagnosis of GBS was made accordingly to clinical and neurophysiological criteria [15]. Based on the nerve conduction study (NCS), the patients were categorized into two groups accordingly to Hadden criteria [15]: acute inflammatory demyelinating polyneuropathy (AIDP), and acute motor axonal neuropathy (AMAN) and acute motor-sensory axonal neuropathy (AMSAN).

In addition, patients with Miller-Fisher syndrome (MFS) were included. The diagnosis of MFS was made on the basis of clinical examination [16]. Antibodies against ganglioside GQ1b were also tested [17].

### Data collection

The following demographic and clinical data were collected:

- Demographics, including age and gender;
- Symptoms at Emergency Department presentation including fever, abdominal pain, vomit, diarrhea, neurological signs and symptoms, preceding infections;
- Data of CSF samples and NCS;
- Treatment of GBS, need for MV or tracheostomy, length of stay (LOS) in hospital, and discharge destination (home or rehabilitation);
- Patients were divided into two groups according to age: <65, ≥65 years. The cut-off of 65 years was chosen according to previous studies of outcome in GBS patients [18,19];
- Comorbidities were defined according to Charlson Comorbidity Index [13];
- mEGOS was calculated for all patients on admission [11].

### Clinical management

Patients diagnosed with GBS were treated, according to guidelines, with supportive medical care and immunotherapy, such as intravenous immunoglobulin (IVIg) and plasma exchange (PE) [4]. Constant ECG and arterial blood pressure monitoring were performed during the hospitalization.

### Study outcomes

In the current study, we compared the values of mEGOS and Charlson Comorbidity Index to determine a correlation between the two tools, and we compared age, values of mEGOS, and Charlson Comorbidity Index between patients discharged to home or rehabilitation and between patients who required MV or not.

Regarding the GBS electrophysiological subtypes, the values of mEGOS and Charlson Comorbidity Index, the type of discharge, and the need for MV were compared in the two subgroups (AIDP and AMAN/AMSAN).

### Statistical analysis

Categorical variables were reported as counts (percentages), while continuous variables (age, LOS, Charlson Index) were reported as median. Categorical variables were compared by Chi-square test, with Fisher's test as appropriate. Continuous variables were compared by Mann–Whitney U-test. A two-sided p-value ≤0.05 was regarded as significant.

Data were analyzed using SPSS for Windows, version 25 (SPSS Inc., Chicago, IL, USA).

## Results

Seventy-eight GBS patients were recruited between 2014 and 2020. Sixty-nine (88.46%) were diagnosed with GBS and nine (11.54%) had classic MFS. An NCS to define the electrophysiological subtypes was available for 43/69 patients. Of 43 patients, 31 patients (72.1%) had AIDP, and 12 patients (27.9%) had AMAN/AMSAN (11 AMAN and 1 ASMAN).

In the patients diagnosed with MFS, antibodies against ganglioside GQ1b were found in 4/9 (44.4%) patients.

Of the 78 GBS patients, 43 (55.13%) were male and 35 (44.87%) female. The mean age was 53.9 (ranged 19–81 years old). Mean LOS in the hospital was 19.29 days (ranged 4–213 days). The main demographic characteristics are summarized in Table 1.

Pure sensory disturbances, including pain, numbness, and paresthesia were found in 33 patients (42.31%) as presenting symptoms. A pure motor disorder at admission was referred in 13 patients (16.67%). Sixteen patients (20.51%) exhibited both weakness and sensory loss at the time of the admission. Five patients (6.41%) complained of diplopia; bulbar symptoms variably associated to sensory symptoms were reported by six patients (7.69%) and two patients (2.56%) presented with respiratory problems (one of those patients presented with dyspnea and dysphagia). Three patients (3.85%) reported facial muscle weakness.

Regarding the CSF findings, we had data available for 43 patients. All patients had cell counts below 50 cells/μL (mean 1–9 range 0–47), and the mean total protein levels were 73.9 mg/dL (range 18–261 mg/dL).

In 35 cases (44.87%), the diagnosis of GBS was suspected by the emergency physicians while in 20 cases (25.64%) by the consultant neurologist in the Emergency Department. Eight patients (10.26%) were transferred from other hospitals. Two patients (2.56%) were referred from general practitioners to our Emergency Department and in four cases (5.13%) patients were sent to the hospital after being evaluated by a neurologist in the outpatient clinic. Nine patients (11.54%) were diagnosed with GBS after admission to the Neurology Department.

**Table 1.** Main demographic characteristics. Abbreviations: *LOS*, length of stay.

| | |
|---|---|
| **Age** | |
| Mean (years) | 53.9 (19–81) |
| **Gender** | |
| Male | 43 (55.13%) |
| Female | 35 (44.87%) |
| **Neurological presentation at admission** | |
| Motor symptoms | 13 (16.67%) |
| Sensory disturbances | 33 (42.31%) |
| Sensory and motor symptoms | 16 (20.51%) |
| Diplopia | 5 (6.41%) |
| Bulbar symptoms | 6 (7.69%) |
| Facial weakness | 3 (3.85%) |
| Respiratory failure | 2 (2.56%) |
| **Symptoms of preceding infection** | |
| Diarrhea | 15 (44.12%) |
| Upper respiratory tract infection | 12 (35.29%) |
| CMV previous documented infection | 1 (2.94%) |
| Fever at admission | 6 (17.65%) |
| **LOS** | |
| Days | 19.29 (4–213). |
| **Treatment** | |
| Plasma Exchange | 7 (8.97 %) |
| IVIg treatment | 58 (74.36%) |
| Not treated | 13 (16.67%) |
| **Discharge destination** | |
| Home | 49 (62.82%) |
| Rehabilitation | 29 (37.18%) |

In our cohort, three patients (3.85%) had autonomic involvement at the time of presentation: two patients had paroxysmal hypertension, and one experienced constipation and urinary retention.

Thirty-four patients (43.59%) had a history of antecedent illness (Table 1).

Sixty-five (83.33%) patients were treated, whereas 13 (16.67%) patients were not treated. The 13 untreated patients had mild GBS (still able to ambulate without assistance). Of the 65 patients treated, 58 (89.23%) received IVIg and 7 (10.77%) PE.

Seven patients (8.97%) received MV and four of those underwent tracheostomy.

On admission mean values for the Charlson Comorbidity Index were 1.20 ± 1.81, ranged 0–12 and the values of mEGOS were 2.4 ± 1.6.

Twenty-nine (37.18%) patients were discharged to outpatient or inpatient rehabilitation and 49 patients (62.82%) with no motor deficits were discharged home. In our cohort, no deaths were reported.

In our study, the rate of home discharge and rehabilitation was similar between elderly and younger patients (p-value =0,969, Cramer's V 0,04).

Tau-b Kendall test was used to measure the correlation between Charlson Comorbidity Index and mEGOS (p-value = 0.005). Comparison of mEGOS and Charlson Comorbidity Index in patients who finally required MV using Mann–Whitney Test showed that those with higher mEGOS received MV, although we did not find a statistical significance between Charlson Comorbidity Index and the need for MV (Mann–Whitney U-test, p-value = 0.009). Similarly, there was no significant difference between the type of discharge and the scores of mEGOS and Charlson Comorbidity Index (p-value = 0.061).

Regarding the electrophysiological subtypes, we did not observe a significant difference in the two subgroups (AIDP and AMAN/AMSAN) concerning the need for MV (p-value = 0.395) and the type of discharge (p-value = 0.248)

Additionally, we did not observe a significant difference of the values of mEGOS (Mann–Whitney U-test, p-value = 0.093) and Charlson Comorbidity Index (Mann–Whitney U-test, p-value = 0.567) in the two electrophysiological subtypes.

## Discussion

GBS is a heterogeneous and often severe disorder. GBS is a commonly missed diagnosis in the emergency department, which significantly increases the morbidity [20], and there is a need for improved treatment and supportive medical care throughout the course of the disease [5].

Regarding signs and symptoms, the results obtained did not differ from those in the literature, being acroparesthesia and mild sensory loss the most common initial symptoms of GBS [16].

Given that GBS presents in the majority of cases with common complaints of mild sensory deficits, it is an easy diagnosis to miss and it should be considered in the Emergency department. Early diagnosis, or at least the suspicion of GBS, is essential to provide a better outcome as delayed therapies can lead to respiratory collapse and life-threatening cardiac arrhythmias. Hence, it is fundamental to emphasize early monitoring for cardiovascular or respiratory failure.

Prognosis and potential predictors of clinical outcome in GBS have been studied extensively [10].

Previous studies suggested that older age was a predictive factor for poor general outcome in GBS patients [8,10]. Other studies demonstrated that older patients have higher mortality; however, the functional recovery may be comparable to younger patients [18].

In the current study, we found no significant differences in age between patients who were discharged home versus patients who needed rehabilitation. This result may imply that age may not influence the short-term prognosis.

mEGOS may be a useful tool for predicting outcomes in patients with GBS and higher mEGOS scores on admission significantly correlated with poor outcomes. This was consistent with the results of a previous study [11].

According to previous studies, the incidence of MV in GBS patients is about 20% [5,7,21]; in our study, this incidence of MV was 8.97%. In our study, patients who ultimately required MV were the ones who had a higher mEGOS score, while the Charlson Comorbidity Index was not related to MV.

Comorbidities were present in the majority of the subjects included in the study, and there was a significant correlation between mEGOS and Charlson Comorbidity Index. Considering the electrophysiological subtypes, we did not find significant differences in the two subgroups for what concerns the need for MV or the type of discharge, and mEGOS and Charlson Comorbidity Index scores were balanced between patients with AIDP when compared with axonal variants. However, we cannot draw conclusions regarding this aspect, because NCS was not available for all the patients and we examined a small cohort.

We certainly know that this study had several limitations, such as being a retrospective analysis and that no deaths were reported in our cohort.

## Conclusion

In summary, age at onset, mEGOS, and Charlson Comorbidity Index scores were well balanced between patients who were discharged to rehabilitation or discharged home. Although there was a correlation between the values of mEGOS and Charlson Comorbidity Index, only patients with higher mEGOS received more frequent MV, confirming the value of this brief score in predicting GBS severity and validating its use even in Emergency settings.

## Disclosure statement

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## Statement of ethics

A general written informed consent to was obtained from all individual participants included in the study. The study was carried out in compliance with the Helsinki Declaration and with the guidelines of the Ethical Committee of our Institution.

Ethical reference number: ID2275.

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