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New Combined Scoring System for Predicting Respiratory Failure in Iraqi Patients with Guillain-Barré Syndrome

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Abstract

Background: The Guillain-Barré syndrome (GBS) is an acute post-infective autoimmune polyradiculoneuropathy, it is the commonest peripheral neuropathy causing respiratory failure. The aim of the study is to use the New Combined Scoring System in anticipating respiratory failure in order to perform elective measures without waiting for emergency situations to occur.

Patients and methods: Fifty patients with GBS were studied. Eight clinical parameters (including progression of patients to maximum weakness, respiratory rate/minute, breath holding count (the number of digits the patient can count in holding his breath), presence of facial muscle weakness (unilateral or bilateral), presence of weakness of the bulbar muscle, weakness of the neck flexor muscle, and limbs weakness) were assessed for each patient and a certain score was given to each parameter, a designed combined score being constructed by taking into consideration all the above mentioned clinical parameters.

Results and discussion: Fifteen patients (30%) that were enrolled in our study developed respiratory failure. There was a highly significant statistical association between the development of respiratory failure and the lower grades of (bulbar muscle weakness score, breath holding count scores, neck muscle weakness score, lower limbs and upper limbs weakness score and respiratory rate score) and above 16 total Combined score(p-value=0.000) .. No significant statistical difference was found regarding the progression to maximum weakness (p-value=0.675) and facial muscle weakness (p-value=0.482).

Conclusion: The patients who obtained a combined score (above 20'30) are at great risk of having respiratory failure.

Keywords: Guillain-Barre Syndrome, respiratory failure, scoring system

1. Introduction

The Guillain-Barré syndrome (GBS) is a group of autoimmune syndromes consisting of segmental demyelination and acute axonal degenerating forms [1]. All GBS variants are a rapidly evolving polyradiculoneuropathy preceded by a triggering event, most often an infection [2]. GBS generally manifests itself as a progressive areflexic weakness with or without autonomic disturbances [(1)]. Its prevalence is between 0·6-4/ 100, 000 per year worldwide [3] and the age ranges from 2 months to 95 years [4], most of the patients being 15- 50 years old [4, 5, 6].

Pathophysiologically, peripheral nerves focal demyelination results in slowing or blocking conduction. Also, there are rare axonal forms [6,7]. 10-30% of (GBS) patients develop ventilatory failure and may require respiratory support. Respiratory failure caused by neuromuscular dysfunctions is particularly dangerous because it develops insidiously [8]. Respiratory failure in GBS is caused by first weakness of the facial, oropharyngeal, and laryngeal muscles. Second weakness of the muscles of inspiration (the diaphragm, intercostals, and accessory muscles) results in inadequate lung expansion and decompensation frequently occurs during night sleep, when the diaphragm effects nearly all the work of breathing [10]. And third expiratory-muscle weakness prevents adequate cough and secretion clearance, increasing the risk of aspiration and pneumonia [10].

The purpose of this study is application and assessment of new combined clinical scoring scale in identifying *patients at risk* of developing respiratory failure in the early phases of the disease, before occurrence of the respiratory failure when elective endotracheal intubation and mechanical ventilation may be performed without waiting for critical situations.

2. Patients and methods

Fifty patients aged between 5-75, with acute GBS, were attended in the neurology ward or ICU in Baghdad Teaching Hospital from May to December-2009. Those patients were examined by neurologists and considered as GBS cases according to the Asbury criteria [11]. There were 25 males (6 were ventilated) and 25 females (9 were ventilated)

Progression to maximum weakness: it is the time to intubation (ventilated patients), or the time to the worst motor function (non ventilated patients) from the onset of symptoms [9]. The patients were divided into 3 groups:

a) progress to peak within 3 days was given 3 points;

b) progress to peak within 4-5 days was given 2 points

c) progress to peak within more than 5 days was given 1 point.

The respiratory rate and digit counting were examined for each patient per minute. Patients are divided into grades as in Table 1

Bulbar weakness, Facial Weakness, and Neck muscle weakness were assessed clinically and graded into groups as in Table 1

Limbs Weakness was assessed according to the Medical Research Council scale [MRC] for muscle power [(12)] and we gave each grade certain points as follows [12].

0 No muscle contraction visible was given 5 points;

1 Flicker of contraction but no movement was given 4 points;

2 Joint movement when effect of gravity eliminated was given 3 points;

3 Movement against gravity but not against examiner's resistance was given 2 points;

4 Movement against resistance but weaker than normal was given 1 point.

5 Normal power was given 0 points. A normal person's total score was 3 out of 30 and the worst score was 30 out of 30

Each patient was examined neurologically and given a certain score according to our scoring system and monitored for signs of respiratory failure.

Statistical evaluation using measurements of central tendency.

Numerical values were evaluated by arithmetic mean (X). While sequential ranking parameters where evaluated by mode. The measurements of dispersion employed were values of the standard deviation (SD) and graphical presentation of the frequency distribution curve. Significant differences between various readings where obtained using an independent test. Statistical parameters and P values were assessed using the standard SPSS (Statistical Package for Social Science) version 17 . [13]

Table [1]. Different clinical parameters grading used in the present study .

A: Progression to maximal weakness:

Time	<3 days	3-5 days	>5 day
Score	3	2	1

B: Respiratory Rate:

Respiratory Rate	≥20/min	15-19	10-14	≤9
Score	1	2	3	4

C: Counting:

Counting	15-20	10-14	≤9
Score	1	2	3

D: Facial Weakness:

Weakness	No	Unilateral	Bilateral
Score	0	1	2

E: Bulbar Weakness:

weakness	No	Mild	Moderate(liquid only)	Severe(only by NGT)
Score	0	1	2	3

F: Neck muscle weakness:

Power Grade	0	1	2	3	4	5
Score	5	4	3	2	1	0

G: Upper Limb Weakness:

Power Grade	0	1	2	3	4	5
Score	5	4	3	2	1	0

H: Lower Limb Weakness:

Power Grade	0	1	2	3	4	5
Score	5	4	3	2	1	0

3. Results

Out of the 50 patients, 50% (25/50) of them were males. The youngest patient in the study was 5 years old and the oldest one was 75 years old.

Fifteen patients needed mechanical ventilation (30%), 9 of them (60%) were females and 6 (40%) were males. The mean of age of the ventilated patients was 40 years old, while the mean for non ventilated patients was 32 years old.

The mean of the combined score for the patients who needed Mechanical ventilation is 19.9/30 and the mean score of patients who did not need MV is 15.8/30. The difference between the two groups of patients is highly significant (p-value=0.000).

The study shows non-significant statistical difference in the progression to maximum weakness between the two groups of patients (ventilated patients (2.5) and non-ventilated patients (2.4) (p-value=0.675). Regarding the facial muscle weakness, this study shows no significant statistical difference between the two groups of patients (p value=0.482) [as seen in Table 2].

The mean score of the respiratory rate, breath-holding count, bulbar muscle weakness, flexor muscles of the neck and limb muscles weakness showed highly significant statistical differences between the two groups of patients (VP &NVP) (the p-value can be seen in Table 2).

In order to obtain the range of scores that represent 95% of the observations that ended in respiratory failure, we used the frequency distribution curve (fig 9) with (2SD) above and below the calculated mean. This gives a value of (16-24) as the limits of interval including the score of patients at risk of developing respiratory failure [see Figure 1].

Table 2: Description and analytical statistics of the two groups of patients (those developing Respiratory failure and those who did not develop Respiratory failure)

	Patient with No RF			Patient with RF			
	Mean	S.D.	Mode	Mean	S.D.	Mode	p value
Progression of maximum weakness	2.4	0.690	3	2.5	0.743	3	0.675

Respiratory rate	2.6	0.497	3	2.1	0.884	3	0.009
Breath holding count	2.1	0.733	2	1.6	0.507	2	0.005
Facial muscle weakness	0.8	0.868	0	1.0	0.926	0	0.482
Bulbar muscle weakness	0.5	0.742	0	2.2	0.676	2	0.000
Neck muscle weakness	2.0	1.485	1	3.2	1.146	4	0.005
Upper limb muscle weakness	2.5	1.291	1	3.5	1.060	3	0.007
Lower limb muscle weakness	2.9	1.089	3	3.9	1.060	3	0.005
Combined score	15.8	3.361	15	19.9	2.264	18	0.000

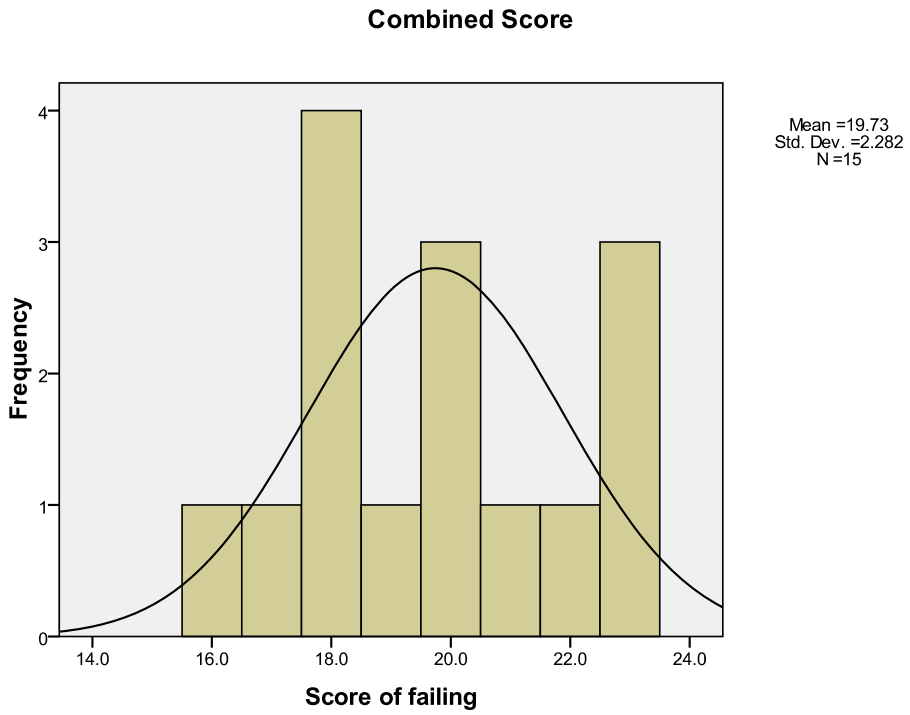

Figure 1: Frequency distribution & bar diagram of the combined score of ventilated patients

4. Discussion

Approximately one third of the hospitalized GBS patients require mechanical ventilation because of respiratory muscle weakness [14-20].

Clinical parameters are advocated to be used to obtain the prognostic profile of the patient, these clinical parameters can easily be evaluated at bedside, they do not need sophisticated procedures and their appraisal will be right away in the hands of attending physicians. Thus, developing clinical prognostic parameters will greatly help to identify patients at risk of respiratory failure and alert the neurologist to take into consideration the possibility of developing RF which is a life-threatening condition, and to take measures to overcome this condition. In order to overcome the disadvantages that are present within each clinical prognostic parameter that has already been mentioned, this study proposes the use of a combined scoring system. This system consists of evaluation of all the previously mentioned clinical parameters, then assigning a particular score for each of them and the sum total of these scoring elements is presented as the combined score of the particular case. The first main advantage of this combined scoring system is the fact that it takes into consideration several clinical parameters and not only the outcome of the particular clinical evaluation, thus, encompassing, in fact, almost all relevant clinical parameters in the evaluation

process. The second advantage is the fact that any disadvantage of a particular clinical parameter will be in fact ameliorated when all parameters are considered all together. Thirdly, this scoring system will yield a numerical value which can be evaluated statistically through statistical methods, can be compared with other studies or observations and correlated with other findings. Above all, the prognostic evaluation parameters of our combined scoring system will tend to be more objective than a single more or less subjective appraisal. Evidence-based medicine is a type of clinical evaluation which means that each clinical entity is treated as "evidence" and the summation of evidences will give the final clinical decision. [21] We divided patients into three groups (less than 3 days progression, between 3-5 days, more than 3 days duration) based on a previous study by Wahab, and other studies [22-25], who found in his study that 70% of patients who had progressed to maximum disability within 3 days needed mechanical ventilation [22-25]. The present study did not detect any significant difference in developing RF between several groups used to evaluate the rapidity of progression (P=0.675). This result is not in agreement with other studies by Lawn et al., [26], Sharshar et al. [27] and Durand et al. [28] who put forward a cutting value of less than one week as a time of duration from the onset to admission as an important clinical predictor of RF. In other words, this work supports the view that what matters is the clinical feature rather than the rapidity of development.

We gave the bifacial muscle weakness 2 points based on evidences from other clinical studies as bilateral facial are more likely to develop respiratory failure [22-26,29].

The present study showed that 11 patients had unilateral facial palsy, 16 bilateral facial palsy, and nevertheless the figures of those patients did not reveal statistical difference when compared with their counterpart according to the possibility of developing respiratory failure. The facial nerve is by all means included in the description of "Bulbar", however it seems that its behavior during the disease entity in GBS is different from other cranial nerves. This is not strange for the facial nerve that shows several discrepancies from other bulbar cranial nerves regarding its anatomical relation of its course and behavior during development, an example of that is the Neurobiotaxis exhibited by this nerve. [20,28]

Bulbar muscle weakness ranks on the top of the clinical parameters with high predictive value of developing respiratory failure in patients with GBS. We graded the presence of bulbar palsy after revision of the results of Wahab. [22], Winer et al., [30], Ropper, et al. [31], Chevrolet et al. [24] and Hughes et al. [25] who found that patients who required mechanical ventilation tended to have bulbar dysfunction.

Evaluation of the respiratory status of the patient is done by several measures, some of them are simple to elicit and evaluate such as the ability of the patient to cough [27] while others are more sophisticated such as using the spirometer to measure the vital capacity [26, 20].

In our study, a "Single-Breath-Count" test is performed by having the patient count out loud after a maximal inspiration. A similar method was also used by in the study of M. Sangeeta. [32] In addition to this method we use the respiratory rate as an independent objective method for evaluation of the respiratory status of the patients.

Evaluation of respiratory rate and single breath count are easier than using a spirometer since it is a clinical bedside method and other measures of inspiratory and expiratory pressures are recommended by Nicholas, et al. [29] that use the famous rule of "20,30,40" (Vital capacity <20ml/kg, Max I pressure < -30 cm H2O, Max E pressure <40cm H2O). [57] Evaluation of the neck flexor muscles is one of the frequently carried neurologic examinations in many neurologic disease states. Many studies attempted to correlate the power of the neck flexor muscles with the possibility of developing respiratory failure. Evidently, this is due to the common site of origin of the innervations of these muscles with the diaphragm at the C4 spinal segment.

Studies showed that weak flexor muscles correlate with developing of respiratory failure [29] or more practically the inability of the patient to lift his head. [19,33]

Our study did not overlook this important parameter and included it in our scoring system of evaluation. However, a more refined scoring is adapted by using Medical Research Counseling Scale (MRCS) of grading to categorize the patients. The result of our work is in agreement with the previously mentioned studies and confirmed the significance of the evaluation of this clinical parameter (P=0.005). This study shows that the type and extent of upper limb motor weakness have a correlation with the possibility of developing respiratory failure (p=0.007). This view contrasts the work of Nicholas, et al. who detected no significant difference in upper limb motor weakness between patients who require mechanical ventilation and those who do not. [29] Other studies are rather vague or imprecise and used the criteria of "inability to flex the arm" as a prognostic indicator of developing RF. Other studies described the inability to lift the elbow as a marker of developing respiratory failure. [29]

Assessment of the power of the lower limbs muscles has also been used for evaluation and the broad term "Inability to stand" has been agreed upon by many studies to be an indicator of poor prognostic factor in those patients and the possibility of developing respiratory failure. [(29)]
In our study, we use the same MRCS grading system to categorize the degree of muscle weakness which is a more accurate and subjective measure. Our study showed statistical significant difference between patients who developed respiratory failure and those who did not (P=0.005).

We hope that these changes in the components of the scoring system and the system itself will tend be a good prognostic marker for predicting the risk of respiratory failure in patients with GBS.

Statistical evaluation showed that there is a highly significant difference between the combined scoring system of patients who developed respiratory failure (mean value about 20) and the combined scoring system of patients who escaped respiratory failure (mean approximately 16).

By using statistical methods for measuring the variability and determination of the standard deviation through a frequency distribution curve (fig 1), it can be concluded that using 2 standard deviations hiatus will encompass 95% of observations, this will give us a value of combined scoring scale range for patients at risk of respiratory failure between (16-24). In other words, patients with a calculated combined score between (16-24) carry a higher statistical risk for developing respiratory failure. This will be correct at a level of significance of 0.05. In our view, this scoring system is a useful mean to evaluate and predict the risk of developing respiratory failure in patients with GBS. We highly advocate its use in clinical setting to get an insight for the possible deterioration in the direction of developing respiratory failure. As with any novel, method further evaluation is by all means needed to give a more solid base for its usefulness, so it is a step in the direction of obtaining a useful answer to our question: "Will this patient develop Respiratory failure?"

5. References

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