

# Comparison between electrophysiological changes and functional gains of patients with Guillain Barré syndrome in the Rehabilitation and Readaptation Center Dr. Henrique Santillo (CRER)

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## ABSTRACT

Progressive acute inflammatory polyradiculopathy, Guillain Barré syndrome (GBS) is commonly associated to a post exposition to an infectious agent or a stimulus, thereby compromising peripheral motor neurons. **Objective:** The objective is to compare electrophysiological changes with functional improvements of GBS on the relation between prognosis and alterations of the electroneuromyography assessment, and to evaluate patients after one year of onset GBS. **Methods:** Retrospective study based on medical reports of the Centro de Reabilitação e Readaptação Dr. Henrique Santillo – CRER of patients registered from 2008 to 2014. **Results:** Forty-eight cases were found, nineteen reports were selected, once they attended the time period criteria. One of these was excluded due to lack of electroneuromyography data, therefore data of eighteen patients were analyzed. **Conclusion:** Rehabilitation is substantial in the final results and in the long term of patients with GBS, whereas rehabilitation program for hospitalized patients is a distinctive work to diminish the losses imposed by GBS, regardless of the functional deficits. The data have shown that the functional improvements acquired one year after GBS onset have no evident relation to what is found in electroneuromyography.

**Keywords:** Guillain-Barre Syndrome, Electromyography, Rehabilitation, Rehabilitation Centers

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## INTRODUCTION

Acute inflammatory polyradiculopathy of progressive characteristics, Guillain Barré syndrome (GBS) is strongly associated with autoimmune factors.<sup>1</sup> It began to be clinically noticed in the eighteenth century, when it had not yet gained such a name, which would arise only in 1916 when Guillain Barré and Strohl described cases of ascending muscular weakness, areflexia, paresthesia and increase of proteins associated to the fall of cellularity in the assessment cerebrospinal fluid.

Considered the main cause of flaccid paralysis in the West, GBS is not predisposed to sex, but studies have shown a slight dominance among males.<sup>1</sup> Usually the condition occurs after exposure to an infectious agent, or a stimulus, that triggers the involvement of peripheral neurons weeks or days after the exposure.

Among the most commonly involved infectious agents are *Campylobacter jejuni*, cytomegalovirus, Epstein Barr and *Mycoplasma pneumoniae*.<sup>2</sup> The pathophysiology of GBS indicates that demyelination varies from focal to extensive, in the presence or absence of cellular infiltration, until the appearance of axonal degeneration, with or without inflammatory infiltrates or demyelination. Multifocal demyelination is the pathological landmark of this disease.<sup>3</sup>

Diagnostic criteria reinforce the importance of knowledge regarding the clinical condition presented by patients with GBS, combined with laboratory examination of cerebrospinal fluid, collected one week after the onset of symptoms, and an electrophysiological study. The clinical scenario is variable, most often starting with a history of paresthesia of the fingers, with ascending muscular weakness, what may affect even cranial pairs, always with total absence or reduction of the deep reflexes.<sup>3</sup>

GBS is considered the most common cause of acute non-traumatic neuromuscular paralysis.<sup>4</sup>

The main subtypes of the disease are Demyelinating Polyradiculopathy, Acute Motor Axonal Neuropathy, Acute Motor-sensory Axonal Neuropathy and Miller Fisher Syndrome. Each one is ranked by the degree of neuronal involvement, manifesting in the most varied ways, which may cause substantial sequelae and even death.<sup>5</sup>

## OBJECTIVE

The objectives of this study is to compare electrophysiological changes and functional gains in GBS, observe the relationship between prognosis and alteration in the electromyography test, and verify the condition of the patients after one year from GBS onset.

## METHODS

This is an observational, retrospective, and longitudinal study, under the ethical principles for research involving human beings of the Brazilian resolution 466/2012 of the National Health Council. This study was approved by the Teaching and Research Commission of the *Centro de Reabilitação e Readaptação Dr. Henrique Santillo – CRER*.

Patients admitted to the CRER between January 1, 2008 and March 31, 2014, with a diagnosis of Guillain Barré Syndrome were selected and evaluated, and the data were collected in a review of medical records at first, and then in direct telephone contact with patients to report the gains after one year of disease onset.

Patients should meet the following inclusion criteria: age, use of immunoglobulin or plasmapheresis, initial and final functional independence measure, length of hospital stay in days, demand mechanical ventilation, electromyography alterations, and health condition one year after the onset of the disease.

For statistical calculations, Kruskal Wallis, U Mann-Whitney test, Chi-square test, Fisher's Exact test were performed with the SPSS Software version 15.0. It was not possible to identify the level of qualification of the physicians who performed the electroneuromyography of the patients, or the protocols they used, once there were no such data in the reports.

## RESULTS

After reviewing medical records of GBS patients treated at the hospital, forty-eight cases were initially identified, of which only nineteen were initially selected once they were admitted according to study period. One patient was excluded because the result of electroneuromyography was not included, and eighteen patients were

included in the analysis. The other patients were excluded because they were outpatients.

The table 1 it shows that patients who presented alterations of the motor-sensory axonal type had a greater variation in the mean of the Functional Independence Measure (FIM), followed by the patients with demyelinating alteration, and finally, those who presented motor axonal alteration. It can also be noticed that regardless the hospital stay, either greater or less than 30 days, had little impact in the FIM variation.

Table 2 shows that patients who had a motor-sensory pattern presented better mean FIM than the others, and that patients older than 60 years of age had higher FIM at the end of hospitalization.

Patients who presented demyelinating and motor-sensory axonal alterations had a predominance in the use of mechanical ventilation (MV) according to Table 3.

Table 4 shows that there was a predominance of lower limb weakness in patients with axonal and motor alterations. It is also noted that even though it was an axonal motor in the electromyography, one patient presented a sensory alteration at physical examination. No patient remained with total limb weakness. The table also shows that in all three types, more than 75% did not need any kind of assistive device for their gait, and that one patient in each type needed a wheelchair for their locomotion. There was one case of death of a patient who had the demyelinating type.

Table 5 shows the changes after one year of disease onset, by analyzing immunoglobulin (IG) at the beginning of the disease. Of the 18 patients analyzed, 16 undertook immunoglobulin treatment. It is noted that even with the use of IG therapy, 8 had lower limb weakness (LL) after one year of onset, regardless the use of IG therapy. It is also possible to observe that only 2 of the patients who received IG therapy presented upper limb impairment. Another observation is that even though they used immunoglobulin, 3 patients needed assistive device for walking and that the use of a wheelchair after one year was very small in patients who used immunoglobulin. Only one case of death was registered one year after GBS.

Table 6 presents the variables studied after one year with the use or not of plasmapheresis, a treatment 6 of the 18 pa-

**Table 1.** Mean, Standard Deviation and Confidence Interval of the FIM difference measured in each observed variable

Observed variable	N	Mean	SD	IC 95%		P
				Inferior	Superior	
<b>ENMG Classification</b>						
1	6	30.00	26.08	2.63	57.37	0.911 <sup>A</sup>
2	4	22.25	19.87	-9.37	53.87	
3	8	30.88	28.14	7.35	54.40	
<b>Hospital stay (days)</b>						
< 30	3	32.67	34.59	-53.25	118.59	0.695 <sup>B</sup>
≥ 30	16	27.31	23.05	15.03	39.59	

A Test: Kruskal Wallis; B Test: U Mann-Whitney; ENMG, electroneuromyography; ENMG classification: 1, Demyelinating; 2, Motor Axonal; 3, Motor-sensory axonal

**Table 2.** Mean, Standard Deviation and Confidence Interval of the FIM difference measured in each observed variable

Observed variable	N	Mean	SD	IC 95%		P
				Inferior	Superior	
<b>ENMG Classification</b>						
1	6	91.67	28.36	61.91	121.43	0.803 <sup>A</sup>
2	4	83.00	25.79	41.96	124.04	
3	8	94.13	31.17	68.07	120.18	
<b>Age (years)</b>						
< 60	15	91.20	30.45	74.34	108.06	0.953 <sup>B</sup>
≥ 60	3	102.00	22.54	46.01	157.99	

A Test: Kruskal Wallis; B Test: U Mann-Whitney. FIM: Functional Independence Measure; ENMG, electroneuromyography; ENMG classification: 1, Demyelinating; 2, Motor Axonal; 3, Motor-sensory axonal

**Table 3.** Number of patients in each analyzed variable according to ENMG classification

Variable	ENMG Classification						P
	1 (N=6)		2 (N=4)		3 (N=8)		
	N	%	n	%	n	%	
<b>Mechanical Ventilation</b>							
Yes	4	66.7	2	50.0	5	62.5	0.606

Test: Chi-square; ENMG classification: 1, Demyelinating; 2, Motor Axonal; 3, Motor-sensory axonal

**Table 4.** Number of patients in each factor analyzed according to ENMG classification

Factor	ENMG Classification						P value
	1 (N=6)		2 (N=4)		3 (N=8)		
	n	%	n	%	n	%	
<b>Weakness LL</b>							
Yes	2	33.3	3	75.0	6	75.0	0.232
<b>Weakness UL</b>							
Yes	2	33.3	1	25.0	1	12.5	0.643
<b>Plegia LL</b>							
No	6	100.0	4	100.0	8	100.0	*
<b>Plegia UL</b>							
NO	6	100.0	4	100.0	8	100.0	*
<b>Sensory alterations</b>							
Yes	2	33.3	1	25.0	5	62.5	0.129
<b>Gait assistive device</b>							
Yes	1	16.7	1	25.0	2	25.0	0.923
<b>Wheelchair</b>							
Yes	1	16.7	1	25.0	1	12.5	0.861
<b>Death</b>							
Yes	1	16.7	—	0.0	—	0.0	0.347

Statistical test: Chi-square; \*As there was no variability, a statistical test was not possible; ENMG, electroneuromyography; ENMG Classification: 1, Demyelinating; 2, Motor Axonal; 3, Motor-sensory axonal; UL, upper limbs; LL, lower limbs

tients undertook. Even with this therapy, 50% of the patients had lower limb weakness one year after disease onset, and only 20% had weakness of upper limbs. Whether or not to apply plasmapheresis therapy did not change the percentage of patients requiring assistive walking devices one year after the disease, as well as the proportion of those who needed a wheelchair for locomotion. The patient who died after one year of the disease had not been treated with plasmapheresis.

## DISCUSSION

Of the many variables analyzed in the study, recovery of muscle strength is the initial goal of patients with GBS. According to Rajabally et al.,<sup>6</sup> on average only 60% of cases have total recovery of muscle strength. Our results show that there was a greater recovery in the upper limb as 14 patients had no upper extremity weakness at the end of one year, whereas 11 patients had some lower limb weakness after one year, especially in patients with demyelinating alterations. There were no plegic patients at the end of the study year. On average, 80% of the patients did not depend on gait assistive device after one year of the disease, with a slight dominance in patients with motor axonal and motor-sensory axonal.

Regarding wheelchair use after one year, there was no significant difference between the subtypes, with the axonal type having a larger domain, 25% of the patients with this alteration needed this means of locomotion.

Our study also revealed that patients with motor-sensory axonal type had greater variation in the FIM, perhaps because they presented greater functional losses, and therefore had chances of obtaining greater gains from the rehabilitation program.

Another relevant finding was the number of patients who depended on mechanical ventilation. Verma et al.<sup>7</sup> show that around 16.7% of the patients the need MV. The CRER study showed that more than 60% of our patients who presented demyelinating and motor-sensory axonal changes in electromyography required mechanical ventilation.

Although axonal motor was shown in the electroneuromyography, 1 patient presented sensitive alteration at physical examination. Capasso et al.<sup>8</sup> report that

**Table 5.** Number of patients in each manifestation after one year of GBS onset, according to the use of IG therapy.

Manifestation	Immunoglobulin				P value
	No (N=2)		Yes (N=16)		
	N	%	N	%	
<b>LL weakness</b>					
Yes	2	100.0	8	50.00	0.485
<b>UL weakness</b>					
Yes	1	50.0	2	12.5	0.386
<b>Sensory changes</b>					
Yes	1	50.0	6	37.5	1.000
<b>Gait assistive device</b>					
Yes	1	50.0	3	18.88	0.386
<b>Wheelchair</b>					
Yes	1	50.0	2	12.5	0.314
<b>Death</b>					
Yes	—	0.0	1	6.25	1.000

Statistical test: Fisher's exact test; UL, upper limbs; LL, lower limbs

**Table 6.** Number of patients in each manifestation after one year of GBS onset, according to plasmapheresis therapy

Manifestation	Plasmapheresis				P
	No (N=12)		Yes (N=6)		
	N	%	N	%	
<b>LL weakness</b>					
Yes	7	58.3	3	50.0	1.000
<b>UL weakness</b>					
Yes	2	20.0	1	20.0	1.000
<b>Plegia LL</b>					
No	12	100.0	6	100.0	*
<b>Plegia UL</b>					
No	12	100.0	6	100.0	*
<b>Sensory changes</b>					
Yes	5	41.7	2	33.3	1.000
<b>Gait assistive device</b>					
Yes	2	20.0	1	20.0	1.000
<b>Wheelchair</b>					
Yes	2	20.0	1	20.0	1.000
<b>Death</b>					
Yes	1	8.3	—	0.0	1.000

Statistical test: Fisher's exact test; \*As there was no variability, a statistical test was not possible; UL, upper limbs; LL, lower limbs

there may be sensory involvement in patients with exclusively axonal changes at the electroneuromyography, although in a smaller amount of fibers in proportion to the motor ones.

Regarding the use of immunoglobulin, the study points out that this therapy yielded no differences for lower limb weakness after one year, as it was found that 50% of those using IG still had some degree of weakness. However, only 18.8% of the

patients used gait assistive device after one year among those who used IG. Perez<sup>5</sup> reports that the use of IG reduces the time the patient needs to perform gait without assistive devices.

Plasmapheresis treatment was given to 6 of the 18 patients. Even with this therapy, 50% of the patients had some weakness in the lower limbs after one year of disease, whereas only one patient presented weakness in the upper limb after this period. Regarding the assistive de-

vices, there was no difference in whether or not to use plasmapheresis therapy, and only 16.7% needed some assistive approach or wheelchair for locomotion. Comparing patients who took immunoglobulin with plasmapheresis at the end of a year, the results are similar to Perez<sup>5</sup> who says that the recovery of patients is similar either using immunoglobulin or using plasmapheresis. He also reports that immunoglobulin has advantages once its administration is easier and it causes less instability to the patient.<sup>5</sup>

## CONCLUSION

After analyzing the data, we concluded that there was no significant difference in the results of patients who used immunoglobulin or plasmapheresis after one year of disease, and that the therapy should be chosen according to the experience of the service or the availability of the treatment.

Physical rehabilitation plays a key role in the outcome and long-term care of patients with GBS, and it is a distinct effort in physical rehabilitation facilities for assuring the capacity to reduce the damages caused by the disease, regardless of the functional deficits.

Associated to the clinic, electroneuromyography is an important tool for the diagnostic, even though its interpretation must be careful, given it is dependent on the operator. Although this assessment have shown subtypes and their probable expected changes, the present study allows us to infer that the functional gains are not directly related to what is found in the electromyography, and that patients who theoretically had greater incapacities, as those with motor-sensory axonal type, for example, they had a greater variation in the functional independence measure (FIM) during the hospitalization period. Also, the general motor alterations along one year of the disease did not have direct correlation with the type shown in the electroneuromyography.

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