

Guillain-Barré syndrome

Pattern of muscle weakness

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ABSTRACT

Objective: The objective of this study was to determine the pattern of muscle weakness in patients with Guillain-Barré Syndrome.

Methods: In a cross-sectional study, 50 Iraqi patients aged one to 60 years diagnosed with Guillain-Barré Syndrome according to Asbury criteria, admitted in 5 Neurological Centers in Baghdad, Iraq between October 1997 and October 1999, were studied for pattern of muscle weakness by clinical evaluation of power using scale from 0-5.

Results: In 80% of patients, muscle weakness started in lower limbs while at presentation 4 limb weakness was the most frequent (96%). It was found that the upper extremity weakness was mainly distal in 73% of patients, while lower extremity weakness was mainly proximal in 68%. Weakness in extremities associated with cranial nerve involvement occurred in 72% of patients. Trunk muscles were involved in 34%. Various modes of spread of muscle weakness were seen in this study but the

ascending variety was the most common occurring in 78% of patients and it was characterized by upward spread, however, contiguous parts of the body were not always successively involved.

Conclusion: Upper extremity weakness was mainly distal while lower extremity weakness was mainly proximal and there is predilection for trunk muscle involvement that is quite unusual in other types of polyneuropathy. Therefore, all patients with suspected Guillain-Barré Syndrome should be examined carefully for pattern of muscle weakness in extremities, which may be helpful in differential diagnosis especially in early stages of the disease.

Keywords: Guillain-Barré syndrome, acute inflammatory, demyelinating polyneuropathy, muscle weakness, polyneuropathy.

Neurosciences 2002; Vol. 7 (3): 176-178

Guillain-Barré syndrome (GBS) is viewed as a reactive, self limited autoimmune disease triggered by a preceding bacterial or viral infection, characterized by rapidly evolving symmetrical limb weakness, loss of tendon reflexes, absent or mild sensory signs and variable autonomic dysfunction.¹ Since the virtual elimination of poliomyelitis, GBS has become the most common cause of acute generalized flaccid paralysis in western countries, with an annual incidence of 0.75-2 cases per 100,000

population.^{1,2} Guillain-Barré syndrome occurs in all parts of the world and in all seasons, it affects children and adults of all ages and both sexes. A mild respiratory or gastrointestinal infection precedes the neurotic symptoms by one to 3 weeks (sometimes longer) in approximately 60-80% of patients.^{3,4} Previous reports suggested that the weakness might be proximal or distal without recognizing any specific pattern. Our study was conducted to confirm an observation noticed during our practice in which

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Received 17th December 2001. Accepted for publication in final form 17th March 2002.

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we found that there is a certain pattern of muscle weakness noticed in many patients presenting with GBS and we hope that it will be a trigger for further studies in the region.

Methods. Fifty patients with GBS were studied between October 1997 and October 1999, their mean age at presentation was 21.1 ± 16.1 years. (range 1-60 years), male to female ratio 3.1:1.

Clinical study design and participants. In a cross sectional study a total of 50 patients with GBS were studied, all were examined by Consultant Neurologists and diagnosed according to Asbury criteria,⁵ admitted in 5 Neurological Centers in Baghdad, Iraq between October 1997 and October 1999.

Clinical monitoring. Motor system was studied in detail including (1) How frequent the muscle weakness is the initial symptom? (2) Pattern of weakness onset involving extremities. (3) Proximal versus distal muscle weakness in extremities. (4) Mode of spread of muscle weakness. (5) Topography of muscle weakness during hospitalization. Strength was recorded using the following scale from 0-5. Zero = no movement, one = flicker or trace of contraction but no associated movement at joints, 2 = movement with gravity eliminated, 3 = movement against gravity but not against resistance, 4- = movement against mild degree of resistance, 4+ = movement against moderate resistance, 5 = full power.

Laboratory methods. Basic laboratory studies and serum sodium and potassium were performed in every patient, throat swab for culture was carried out in 29 patients, pulmonary function test was carried out in 33 patients, nerve conduction measurement and electromyogram (EMG) were carried out for 45 patients, cerebrospinal fluid (CSF) aspiration was carried out for 37 patients and samples of CSF were studied for sugar, protein and cells and in some case for Rose-Bengal test.

Statistical methods. Statistical analysis was carried out according to student's t-test for difference between 2 means. P value <0.05 was considered statistically significant.

Results. Most of the cases reported during the cold period of the year. Muscle weakness was the initial symptom in 68% of patients and in 80% muscle weakness started in lower limbs (**Tables 1 & 2**). At time of presentation, motor manifestation was present in all patients involving the extremities with or without cranial nerves involvement. Four extremity muscle weakness were the most frequent, present in 96% of patients, and it was found that the upper extremity weakness was mainly distal in 73% of patients, while lower extremity weakness was mainly proximal in 68% of patients (**Table 3**).

Table 1 - Symptoms of 50 Guillain-Barré syndrome patients.

Symptoms	Initial	At admission
	N (%)	N (%)
Motor weakness	34 (68)	50 (100)
Alone	17 (34)	17 (34)
With parasthesia	8 (16)	14 (28)
With pain	4 (8)	6 (12)
With pain and parasthesia	5 (10)	13 (26)
Sensory complaints	16 (32)	0 (0)
Pain alone	2 (4)	0 (0)
Parasthesia alone	6 (12)	0 (0)
Pain and parasthesia	8 (16)	0 (0)
Total	50	50
N - number		

Table 2 - Pattern of weakness onset involving extremities.

Beginning of weakness	N (%)
UL	8 (16)
LL	40 (80)
Simultaneous UL and LL	2 (4)
Total	50 (100)
N - number, UL - upper limb, LL - lower limb.	

Table 3 - Proximal versus distal motor weakness in the extremities.

Weakness	UL	LL
	N (%)	N (%)
Greater proximal	5 (10.4)	34 (68)
Greater distal	35 (72.9)	6 (12)
Equal	8 (16.7)	10 (20)
Total	48 (100)	50 (100)
N - number, UL - upper limb, LL - lower limb		

Weakness in extremities was associated with cranial nerves involvement or trunk muscle paralysis occurring in 72% of patients, while in 28% of the patients weakness involved the extremities only. Various modes of spread of muscle weakness were seen in this study. The ascending variety was the most common, occurring in 78% of patients. This was characterized by upward spread, but contiguous parts of the body were not always successively involved. In addition the patients had one or more of these features; 32 had bilateral facial weakness, 21 trunk muscle paralysis, 20 bulbar weakness, 2 extra ocular muscle weakness and 2 had 5th nerve weakness. The mean duration of time interval from the onset of illness to severe deficit using tetraplegia as criterion for severity was 8.88 days \pm 4.75 SD (range 3-23 days) for those who recovered and 4.25 days \pm 4.06 SD (range 1-11 days), for those who died the relation between mortality and time interval from onset to tetraplegia proves to have statistically significant correlation (P 0.015).

Discussion. Apart from yearly summer epidemics among rural children in northern China,^{1,6} most studies show only minor seasonal variations.^{2,7,8} While in our study a clustering of patients is noted during the cold period of the year (75%). Previous reports consider antecedent diarrhea illness as a bad prognostic point,^{1,9} while in our study 2 patients out of 8 developed respiratory paralysis, both were recovered. Weakness at the start of symptoms is more commonly involves the lower limbs (80%), while at time of presentation motor manifestations were present in extremities with or without cranial nerve involvement and 4 extremities weakness was the most frequent presentation (96%), which is in agreement with previous reports.^{1-3,10} Osler and Sidell¹¹ emphasized that proximal weakness is more frequent than distal, while Masucci and Kurtzke,¹² Loffel et al¹³ and Marshal⁸ suggested that the weakness might be proximal or distal without recognizing any specific pattern. In our patients, we found that the upper extremity weakness was mainly distal while lower extremity weakness was mainly proximal, and this pattern of weakness may help in the differential diagnosis especially at an early stage of the disease. Osler and Sidell¹¹ stated that severe involvement of the trunk muscles is uncommon. The trunk muscles were involved in 34% of our patients, often severely, and in all the 4 limbs were also weak which is in agreement with Masucci and Kurtzke¹² and Marshal's series.⁸ Trunk weakness in a patient with progressive polyneuropathy favors the diagnosis

of GBS as it is an uncommon finding in most polyneuropathies due to other causes. We found that cranial nerves were involved in 68% of patients and the facial nerve (bilateral) was the most frequently affected (64%) and next were the 9th and 10th cranial nerves (40%). This is in agreement with previous reports.¹⁻⁵ We think that the ascending variety of spread of muscle weakness was overemphasized since other forms of spread could occur also, which is in agreement with Masucci and Kurtzke.¹² Relation of mortality to time interval from onset to severe deficit was statistically significant (P=0.015). This is consistent with previous reports, which showed that rapid progression of disease is a bad prognostic point.^{1,2,14}

In conclusion, the patients with GBS upper extremity weakness was mainly distal while lower extremity weakness was mainly proximal and there is predilection for trunk muscle involvement that is quite unusual in other types of polyneuropathy. Therefore, all patients with GBS should be examined carefully for pattern of muscle weakness in extremities which may be helpful in the differential diagnosis especially at an early stage of the disease.

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