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RESPIRATORY DISTRESS IN PATIENTS OF GUILLAIN-BARRE SYNDROME WITH SENSORY MANIFESTATIONS

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ABSTRACT:

INTRODUCTION: Guillain-Barré syndrome (GBS) is one of the most frequent disorder of peripheral nervous system associated with respiratory distress. One clinical parameter that was found significant in heralding the onset of respiratory failure in GBS weresensory manifestations. The aim of the study was to determine the frequency of respiratory distress in patients of GuillainBarresyndrome with or without sensory manifestations in the disease. **MATERIALS AND METHODS:** This was a descriptive case seriesthat was conducted from January 19, 2013 to July 18, 2013 in Department of Neurology, Pakistan Institute of Medical Sciences, Islamabad. All patients who met the inclusion criteria were selected for the study. Sensory manifestations of GBS were determined clinically, and patients were divided into two groupsi.e patients with sensory manifestations and without sensory manifestations. These two groups were followed for a period of 10 days and monitored for the presence of respiratory distress. **RESULTS:** A total of 110 patients were included. The mean age of patients was 34.41 years with standard deviation of 17.12 years. 76 (69.09%) patients were male and 34 (30.91%) patients were female. 85 patients had sensory manifestations while 25 patients had no sensory manifestations. 33 patients of GBS with sensory manifestation had respiratory distress while 52 patients had no respiratory distress.**CONCLUSION:** In our study moderate to severe pain is a common and early symptom of Guillain-Barre Syndrome. Respiratory involvement is more common in patients with sensory manifestation. Early detection and aggressive treatment of sensory manifestations of GuillainBarresyndrome should be a part of initial management so as to minimize the risk of respiratory failure.

INTRODUCTION:

Guillain-Barre syndrome is an acute, post-infectious, autoimmune disease involving the demyelination of peripheral nerves leading to loss of nerve signaling and symmetrical ascending paralysis. Symptoms of a preceding upper respiratory or gastrointestinal tract infection are present. Campylobacter jejuni, Cytomegalovirus, Epstein barr virus and mycoplasma pneumonia are the predominant causes.1-3These infectious agents may induce antibody production against gangliosides and glycolipids of myelin in the peripheral nerves. The annual incidence varies from 0.4 to 4.0 cases per 100,000 persons with a median incidence of 2 cases per 100,000 population.[4,5]

Guillain Barré syndrome (GBS) is one of the most frequent disorder of peripheral nervous system associated with respiratory distress.1 At presentation, about 40% patients have respiratory weakness and respiratory failure requiringventilatory support occurs in up to one third of patients at some time during the course of their disease.[6] Sensory symptoms are paresthesias and numbness in toes and fingers, more than half of patients complain of aches and pains in muscles, few patients have burning sensations in fingers and toes, by the end of first week of onset, vibration and joint position sense in fingers and toes are reduced, deep sensibility (touch, pressure, vibration) are more affected than superficial sensations (pain, temperature)[7,8].Few studies have demonstrated the association of sensory manifestations of GuillainBarresyndrome with respiratory distress[9] but so far to our knowledge this topic is not studied locally in our setup. The aim of the study was to determine and compare the frequency of respiratory distress in patients of GuillainBarre Syndrome who present with and without sensory manifestations in our population so as to aid in early detection of onset of respiratory failure in the affected individuals.

MATERIALS AND METHODS:

This was a descriptive case study conducted in the Department of Neurology, Pakistan Institute of Medical Sciences,Islamabad for six months fromJanuary 19, 2013 to July 18, 2013 after taking permission from ethical committee of hospital.A total of 110 patients of GuillainBarresyndrome were enrolled using non probability consecutive sampling. Sample size was calculated using WHO sample size calculator. An informed written consent was taken from all the patients. All patients of both genders above 13 years of age,with GuillainBarre Syndrome (diagnosed on basis of clinical and electrophysiological criteria) were enrolled in this study.Diagnostic criteria for GBS was defined from the NationalInstitute of Neurological Disorders and Stroke (NINDS).[9] According to which the requiredfeatures for the diagnosis of GBS are: Progressive weakness of more than one limb andareflexia. While Supportive features (If any two of the following are present) are:Progression of symptoms over days to four weeks, Relative symmetry, sensory signs/symptoms, Cranial nerve involvement, Autonomic dysfunction, CSF protein >45 mg/dl, cell count <10 mm3.Electrodiagnostic abnormalities for diagnosis of demyelinating variant of GBS (If any two of the following four are present): are, Prolonged distal latencies in two or more nerves, Slowing of Conduction Velocity in two or more nerve,Prolonged F response in one or more nerves, Conduction block in one or more nerves.[10] While for the axonal variants of Guillainbarre syndrome criteria was set as no evidence of demyelination, but reduced amplitudes of compound muscle action potentials (CMAP) in two or more motor nerves in AMAN while for AMSAN, in addition to the criteria set for AMAN(i.e no evidence of demyelination and reduced amplitudes of motor CMAPs), reduction of sensory nerve action potentials (SNAPs) in two or more nerves. For Miller fisher variant reduced or absent SNAPs without involvement of motor nerves and no evidence of demylination.11Sensory manifestations were determined clinically if patients complained of muscle pains and aches,paresthesias (burning, tingling, pins and needles sensations), numbness in fingers and toes, impaired pinprick , joint position and vibration sensations. Respiratory distress was defined as if single breath count is less than 20.[7]

Those patients who had a sensory level, having systemic illness other than Guillain-Barre Syndrome associated with neuropathy that could lead to chronic sensory symptoms on historical inquiry e.g. Diabetes mellitus, chronic renal failure, hereditary neuropathies and toxin exposure,use of drugs causing peripheral neuropathy e.gAntituberculous drugs, Metronidazole, Amiodarone, Phenytoin, Lithium and Hypokalemic paralysis (serum < 3.0 mmol/lit) were excluded. Patients who fulfilled the criteria underwent detailed history and neurological examination. Patients were divided into two groups i.e. patients with sensory manifestations and without sensory manifestations, these two groups was followed for a period of 10 days and monitored for the presence of respiratory distress. The data was entered on a standardized Performa. Data was entered and analyzed using SPSS version 17. Mean and standard deviation was calculated for numerical variables (age). Frequencies and percentages were calculated for categorical variables (gender, sensory signs and symptoms and respiratory distress).

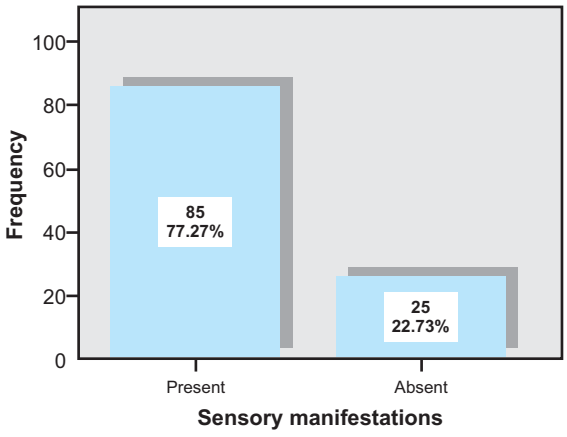
Chi square test was used to compare the presence of respiratory distress between the two groups. P value < 0.05 was considered significant for all the results.

RESULTS:

A total of 110 patients were included in this study. The mean age of patients was 34.41±17.12.Out of 110 patients, 76 (69.09%) patients were male and 34 (30.91%) patients were females.

85 patients had sensory manifestations while 25 patients had no sensory manifestations as shown in figure no: 1

Figure no: 1
Sensory manifestations in different patients



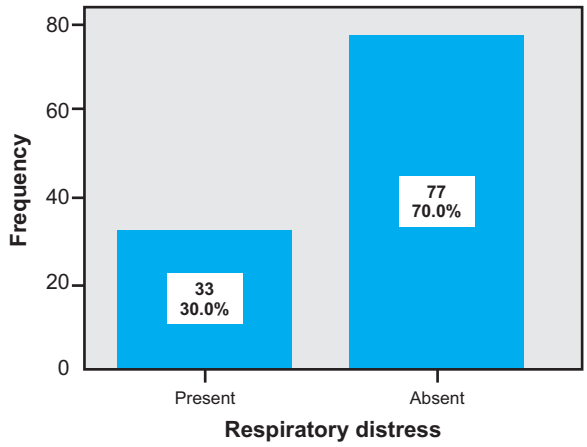
55 male patients had sensory manifestation while 21 patients had no sensory manifestations and 30 female patients had sensory manifestations while 4 patients had no sensory manifestations with insignificant p value of 0.066 as shown in table no: 1.
There was no significant age predilection

Table no: 1
Sensory manifestations in different gender of patients

Gender of patients	Sensory manifestations		Total	p-value
	Present	Absent		
Male	**55 72.4%**	**21 27.6%**	**76 100.0%**	0.066
Female	**30 88.2%**	**4 11.8%**	**34 100.0%**	
Total	**85 77.3%**	**25 22.7%**	**110 100.0%**	

Out of 110 patients, 33 patients had respiratory distress while 77 patients had no
respiratory distress as shown in figure no: 2

Figure no:2
Frequency of respiratory distress in different patients



24 male patients had respiratory distress while 52 patients had no respiratory distress
and 9 female patients had respiratory distress while 25 patients had no respiratory
distress with insignificant p value of 0.589 as shown in table no: 2 while there was no significant age predilection.

Table no: 2
Respiratory distress in different gender of patients

Gender of patients	Respiratory distress		Total	p-value
	Present	Absent		
Male	**24 72.4%**	**52 27.6%**	**76 100.0%**	0.589
Female	**9 88.2%**	**25 11.8%**	**34 100.0%**	
Total	**33 77.3%**	**77 22.7%**	**110 100.0%**	

33 patients of GBS with sensory manifestation had respiratory distress while 52 patients had no respiratory distress and 25 patients of GBS without sensory manifestation had no respiratory distress with significant p value of 0.001 as shown in table no: 3

Table no: 3
Respiratory distress in patients with and without sensory manifestations

		Respiratory distress		Total	p-value
Sensory manifestations	Present	**33 38.8%**	**52 61.2%**	**85 100.0%**	**0.001**
	Absent	**0 0%**	**25 100.0%**	**25 100.0%**	
Total		**33 30.0%**	**77 70.0%**	**110 100.0%**	

DISCUSSION:

GB syndrome is one of the severe forms of polyneuropathies with variable sensory symptoms, motor disability, need for hospitalization and uncertainty of the course of the disease. Sensory disturbances, especially pain, are common in GBS.[12]

The major reason for mechanical ventilation is respiratory muscle weakness and inability to clear secretions.These symptoms often follow ascending extremity weakness but can appear with minimal limb involvement or uncommonly as the sole presenting feature of illness. The pace of progression of respiratory failure from onset can be dramatic, necessitating the need for ventilator support over 24 to 48 hours. Alternatively the progression can be sub-acute, or progressing gradually over 3 to 4 weeks.[13] A careful examination including single breath count, along with arterial blood gases and bed side pulmonary function tests are of extreme importance.14The general recommendation is to intubate these patients when the vital capacity falls to approximately 15 mL/kg.[15] Early predictors of need for mechanical ventilation includedifficulty in clearing secretions because of inability to cough, inability to lift elbows and headand bulbar weakness because of resultant lack of oropharyngeal protective mechanisms.[15,16] Literature has many studies showing the prevalence of sensory symptoms predominately pain in patients with GuillainBarre syndrome in the acute phase of illness and its impact on long term outcome in these patients[17,18], However,

there are only few focused studies in the literature about the association of these sensory manifestations with respiratory distress in patients of GuilainBarre syndrome. So more focus is required to detect the sensory symptoms for early prediction of the heralding respiratory failure in these patients.In our study, out of 110 patients the frequency of sensory manifestation was 77.27% and these manifestations were absent in 22.73% patients.33 patients (30.0%) had respiratory distress. 76 (69.09%) patients were male and 34 (30.91%) were female. The mean age was 34.41±17.12. On comparing various parameters among patients with GBS with and without sensory manifestations, the only statistically significant difference was the presence of respiratory involvement in those with sensory manifestations. None of the other factors or predilection of these sensory symptoms and respiratory distress for age or gender were significant.These results were comparable to the results of other international studies. Karkare et al studied a cohort of 60 patients of GuillainBarre syndrome and found sensory dysfunction in 48 (80%) patients, pain in 30 (50%), paresthesias in 45 (75%), sensation of pain and temp, joint position and vibration were impaired in 8 (13.3%), 14 (23.3%) and 11 (18.3%) patients respectively. Pain assessment using VAPAR(visual analogue for paresthesias), VAP (visual analogue for pain) and VERP (verbal rating scale for pain) from Day 1 to Day 10 of hospitalization was done. On comparing various parameters among patients of GuillainBarre syndrome with (39 patients) and without pain and paresthesias(21patients), statistically significant difference was presence of respiratory involvement in 8 patients (20.5 % and p value = 0.02) with pain and paresthesias and in none of the patients with out sensory menifestations.[9] A study conducted by Taly AB et al[19] showed that sensory symptoms or signs were present in 45% and 59% of patients of GBS in upper and lower limbs respectively and were distal and symmetrical. Impairment of joint position and vibration sense was the commonest finding and was associated with a greater need for ventilatory support and autonomic disturbances.Ruts et al[20] recently described 156 patients with GB syndrome and pain was reported 2 weeks preceding weakness in 36% of patients, 66% reported pain in the acute phase (first 3 weeks after inclusion), and 38% reported pain after 1 year. In the majority of patients, the intensity of pain was moderate to severe. Longitudinal analysis showed high mean pain intensity scores during the entire follow-up. Pain occurred in the whole spectrum of GBS. The mean pain intensity was predominantly high in patients with GBS, patients with sensory disturbances, and severely affected patients. Only during later stages of disease, severity of weakness and disability were significantly correlated with intensity of pain.Walling AD et al[21] showed that the most common form of the disease, acute inflammatory demyelinating polyradiculoneuropathy, presents as progressive motor weakness, usually beginning in the legs and advancing proximally. Symptoms typically peak within four weeks, then plateau before resolving. More than one-half of patients experience severe pain, and about two-thirds have autonomic symptoms, such as cardiac arrhythmias, blood pressure instability, or urinary retention. Advancing symptoms may compromise respiration and vital functions.In the current study, several limitations need to be recognized. The patients recruited were not critically ill as this might have hampered their participation in a questionnaire-based enquiry. Another limitation was non-consecutive selection of subjects depending on the feasibility of evaluation by the investigator. Further, patients were admitted on different days of their illness as is observed in the majority of the studies involving GBS. The electrophysiological studies were carried out early and on variable days of hospitalization.

CONCLUSION:

Sensory manifestations in Guillain-Barre Syndrome are often under-recognized and under-emphasized. Respiratory involvement is more common in patients with sensory manifestation than in those without sensory manifestations.These symptoms require early detection to categorize the patients who are at risk of respiratory failure and hence aggressively managed so as to improve symptomatic profile and quality of life.

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Maher Bano Malik; data collection, data analysis, manuscript writing, manuscript review
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Jahangir Shoro; data analysis, manuscript writing, manuscript review