European Journal of Molecular & Clinical Medicine

ISSN 2515-8260 Volume 09, Issue 06, 2022

ORIGINAL RESEARCH

Pattern of GBS in Kashmir, a Northern Region in India

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ABSTRACT

Background: Guillain-Barré syndrome (GBS) is said to be most common cause of acute flaccid paralysis worldwide. It is also one of the important life-threatening emergencies requiring critical cares in neurology. There are many subtypes of GBS like acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonopathy (AMAN) and acute motor sensoryaxonopathy (AMSAN). GBS shows a lot of variation in the demographic variables like gender distribution and seasonal variation as seen in different studies. The frequency of different variants of GBS is also quite variable. AIDP is more common in Europe and North America whereas axonal subtypes are more often seen in Asia and South America. We conducted this study over a period of Two years to determine these features of GBS in our population.

Material and Methods: 92 patients comprising of 49 male and 43 female patients were included in this study. All these patients were admitted in our hospital and were evaluated as per the established protocol. Baseline investigations were done in all the patients. Neurophysiology and CSF analysis was also done.

Results: In our study we found that most of the patients had AIDP. A preceding precipitating event was present in 31 patients. There was a significant seasonal variation in our population with majority of the cases occurring in spring season. We also found that there were a significant number of patients who presented to us in postpartum period.

Conclusion: This study shows that knowing the demographic variations of GBS patients of a region is important so as to prepare the healthcare facilities for better management of such cases.

Keywords: GBS, Neuropathy.

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INTRODUCTION

The term demyelination describes a loss of myelin with relative preservation of axons. This results from diseases that damage myelin sheaths or the cells that form them. Central nervous system myelin and peripheral nervous system myelin are antigenically different as befits the fact that CNS myelin comes from oligodendroglia and PNS myelin comes from Schwann cells. Therefore, some demyelinating disorders attack the central nervous system like multiple sclerosis, while others affect the peripheral nervous system, the prototype being Guillain-Barre syndrome (GBS). GBS is an acute onset peripheral neuropathy with a rapidly

developing motor weakness and sensory symptoms.[1,2,3] It is diagnosed as per Ashbury criteria,[2] though now a new set of criteria have been suggested.[4] GBS causes respiratory failure requiring ventilator support in approximately 25% with a mortality rate of 4-15%. The most frequent subtype of GBS in North America and Europe is acute inflammatory demyelinating polyradiculoneuropathy (AIDP) whereas in Asia, South America, and Central America, the axonal variants of GBS [Acute motor axonopathy (AMAN) and Acute motor sensoryaxonopathy (AMSAN)] are more common. The treatment options for GBS include plasma exchange which was found to be effective in 1980's and intravenous immunoglobulin (IVIG) whose efficacy was demonstrated in 1990's. Most studies done to compare the two treatment options have shown equal efficacy. The American Academy of Neurology practice guidelines recommend Either IVIG or plasmapheresis for GBS patients with severe disease who have restricted mobility. IVIG is more expensive as compared to plasmapheresis, however it is easier to administer and is safer in patients with autonomic disturbances. The epidemiology and demographic features of GBS are quite variable in different continents as shown by various studies. This study was done to know the features of GBS in a northern state of India.

MATERIAL AND METHODS

This study was conducted by a tertiary care hospital, Government Medical College Srinagar in Jammu and Kashmir State of India. All the patients of GBS admitted by the department of Medicine from October 2020 to September 2022 were included in this study. Data for the study was collected through personal interviews of patients, review of medical records, physical and neurological examination done by a physician and a neurologist. GBS was diagnosed as per standard international criteria.[2,4] In accordance with the Ashbury criteria all these patients had a history of acute ascending weakness of limbs with areflexia. Nerve conduction test with evaluation of median, ulnar, common peroneal, tibial and sural nerves was done in all these patients to document the type of neuropathy.[5] Electrophysiological examinations were performed within 3 weeks of the onset of illness in all patients. CSF analysis was also done in all the patients in the second week of their illness, after taking a proper consent for the procedure. Further sub-classification of GBS into subtypes like AIDP, AMAN and AMSAN, was done on the basis of clinical and Electrophysiological criteria.

RESULTS

The total number of GBS patients admitted in the study period was 92 which included 49 males and 43 female patients. The age distribution and other parameters of these patients are shown in table
1. As shown in this table, the majority of patients were in 2nd and 3rd decade.

Table 1: Variables of GBS patients

Age Group (Yrs)	**Number of Patients (92)**
18 – 30	41
31 – 40	26
41 – 50	12
> 51	13
Type of GBS	
AIDP	71
AMSAN	10
AMAN	5
RECURRENT AIDP	3
POLYNEURITIS CRANIALIS	2

MFS	1
Preceding Event	
Post-partum period	14
Respiratory tract infection	13
pregnancy	2
Gastroenteritis	2
Duration of Hospitalization (Weeks)	
One	56
Two	30
Three	4
Four	2

Different varients of GBS that were seen during the study period are shown in [Figure 1] and as can be seen the most common variant was AIDP.

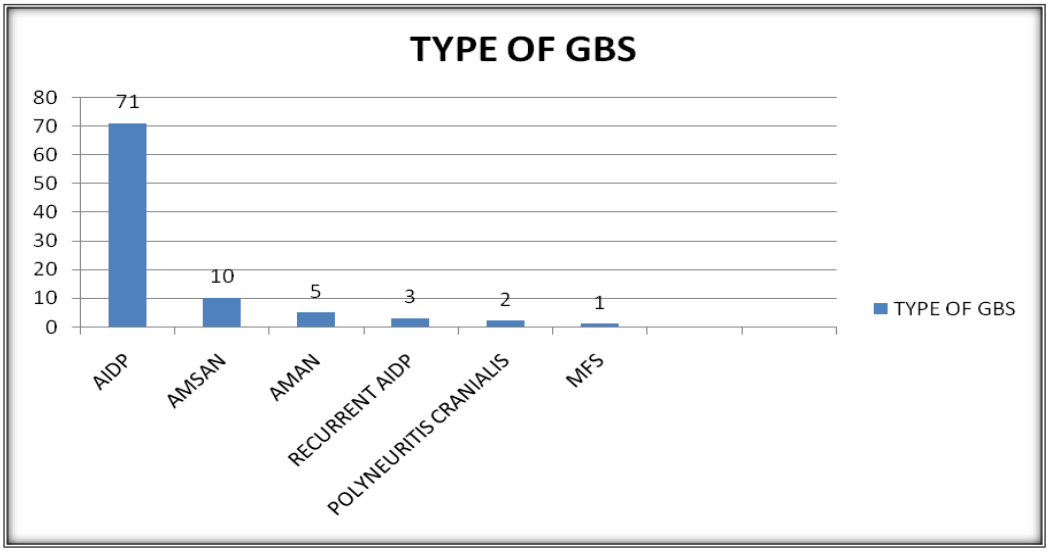

Figure 1: Varients of GBS seen during the study period.

A preceding event like infection and pregnancy was present in 31 patients as shown in figure 2 below.

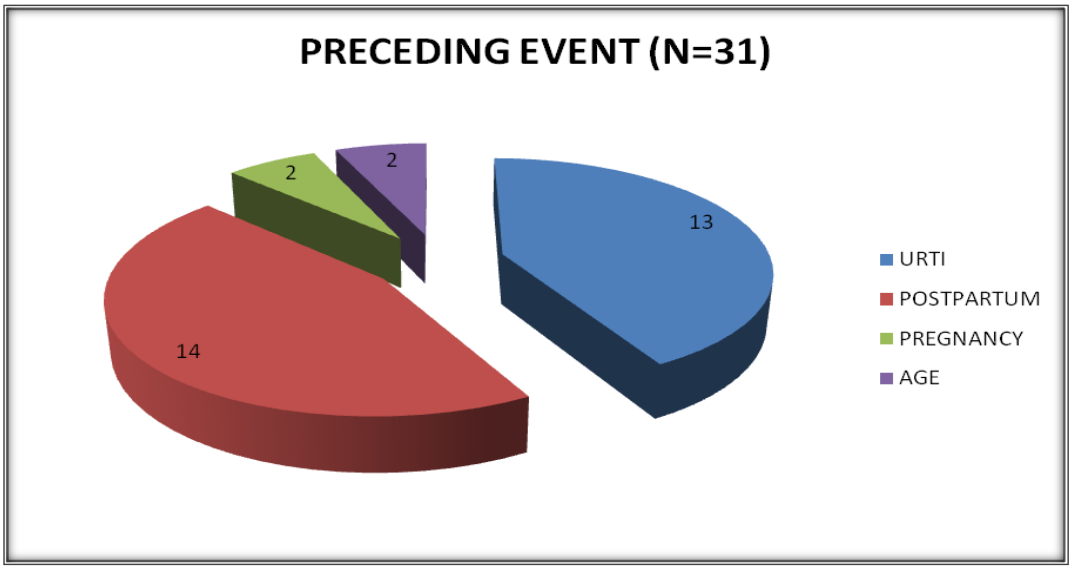

Figure 2: Pie-chart showing the presence of a preceding event in GBS patients.

Out of 14 patients that presented in postpartum period, 8 had undergone LSCS and 6 had a normal Delivery. Seasonal variation of GBS was found which showed that maximum number of cases occurred in the months of March, April and May as has been depicted in figure 3. 5 patients out of 92 have dysautonomia.4 patients needed ventilator support. Only 5 patients received IVIG and 4 patients received Plasmapheresis.

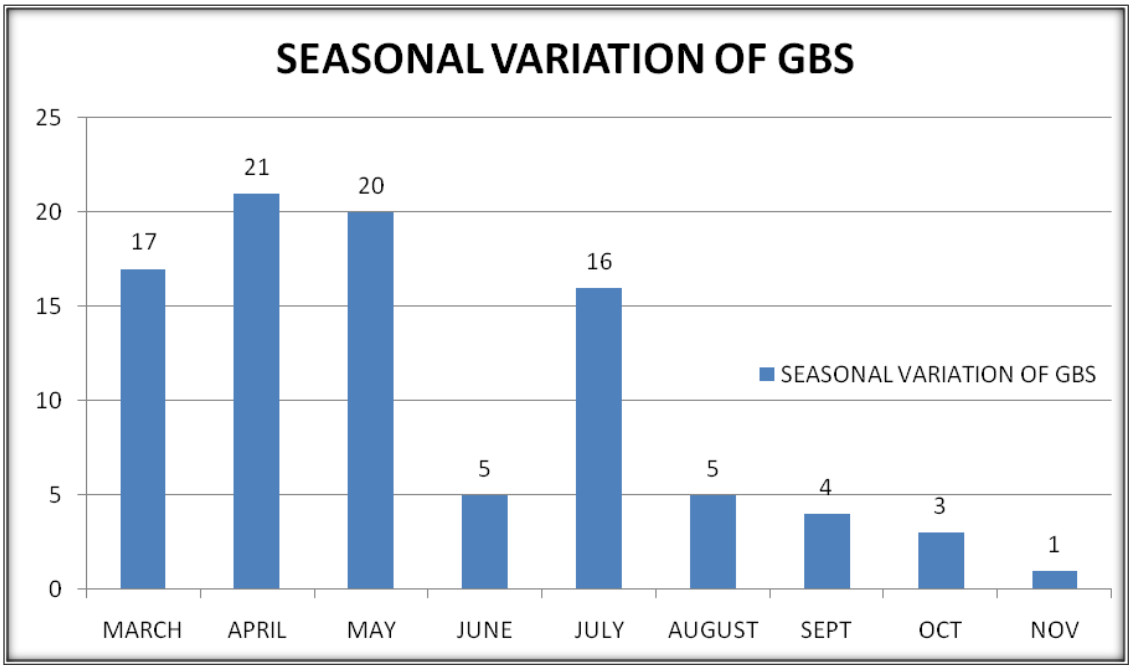

Figure 3: Bar chart showing the seasonal variation of GBS in the study period.

DISCUSSION

GBS is said to be the most common cause of acute flaccid paralysis globally.[6,7,8] The disease presents as an acute inflammatory polyradiculoneuropathy with diminished reflexes and resultant weakness. Most of the patients complain of paresthesias, numbness, or similar sensory changes. Paresthesia begins in the toes and fingertips, progressing upwards, but generally does not extend beyond the wrists or ankles. Pain is usually present in GBS which is most severe in the shoulder girdle, back, buttocks and thighs and occurs even with the slightest movements. The pain in GBS is often described as throbbing or aching in nature.

The median incidence of GBS has been estimated to be 1.11 per 100,000 person-years in the developed countries. The male and female gender ratio has been observed to be 1.78.[6] However in our study the ratio was only 1.14. Nearly two-thirds of cases are preceded by symptoms of upper respiratory tract infection or diarrhea and rarely vaccination (e.g. A/ New Jersey/1976/H1N1 "swine flu" vaccine). In our study a preceding event was found in only one-third of cases which is quite less. We found that thirteen patients had a respiratory tract infection and only two had acute gastroenteritis. The exact reason for having a low incidence of preceding event could not be established. One explanation could be that there was a very mild illness or an asymptomatic viral infection which was responsible for precipitating GBS. This would also mean that our population is more prone to immune-mediated disorders due to some genetic predisposition. Further studies will be needed to determine the exact reason. Among the electrophysiological subtypes, AIDP is considered to be the most common in western countries; whereas, axonal types are more common in Asian countries.[6,7,8] However, studies from India have yielded varying results.[9,10,11,12] Though many studies reveal that the most common variant is AIDP,[9,10,11] but some have shown the axonal variant to be more common.[12] Our study also revealed that the most common variant of GBS in our state is AIDP followed by AMSAN and AMAN varients. Other subtypes of GBS were rare. Miller Fisher Syndrome (MFS), also called Fisher's syndrome is named after Dr. C. Miller Fisher who described it in 1956. The characteristic triad of MFS consist of ataxia, areflexia and opthalmoplegia. It is a rare variant of GBS, observed in only about 1% to 5% of all cases of GBS in Western countries. Miller Fisher syndrome occurs in more men than women by a ratio of approximately 2:1.The mean age of onset of MFS is 43.6 years,though onset has been documented in individuals between the ages of 13 and 78 years. Anti-GQ1b antibodies, activated by certain strains of Campylobacter jejuni, have a relatively high specificity and sensitivity for MFS. Dense concentrations of GQ1b ganglioside are found in the oculomotor nerve (cranial nerve III), trochlear nerve (cranial nerve IV), and abducens nerve (cranial nerve VI) of patients with MFS, which may explain the relationship between anti-GQ1b antibodies and ophthalmoplegia. Titers of anti-GQ1b antibodies in CSF that are greater than 1:40 are specific for MFS. So these antibodies can help in diagnosing MFS especially in those cases were only one or two of the characterisitic triad is present. In our study we found only one case of Miller-fisher syndrome. The risk of GBS is said to decrease during pregnancy and increase in postpartum period. This was true in our study also as 14 patients were in postpartum period but none was seen during pregnancy. The relation between GBS and pregnancy /postpartum period has been studied since long. GBS complicating pregnancy is a rare condition with an incidence of 1.2 and 1.9 cases per 100,000 annually, with a high maternal risk.[13] One of the studies done in Sweden,[14] found that the risk of GBS increases in postpartum period. They reported 21 cases of GBS during pregnancy AND postpartum period over a period of 15 years. However in our series there were 14 cases in only two years period. It has been postulated that cell mediated immunity plays a major role in the pathogenesis and the worsening in postpartum period to be due to a rapid increase in delayed type hypersensitivity during this period. The exact risk of relapse or recurrence in subsequent pregnancies is not known though relapse during successive pregnancies has been reported.[15] A favourable outcome with full recovery has been seen in 70-80% of patients of treated patients. GBS occurring in pregnancy is associated with an increased need for ventilator support, and an increase in maternal mortality up to 7% and upto 20% patients are disabled even after a period of 1 year. Seasonal variation in incidence of GBS is quite well known. One study from Iran,[16] has shown that Spring and winter had the most amount of patients, with admissions from the month of February through June inclusive accounting for 50% of all cases. Studies from India,[17,18] have shown variable seasonal variation with some showing more cases in summer and some in spring season. Our study is somewhat similar to the study

from Iran,[16] as most of the patients were present in first few months of the year. The reason behind this strong seasonal variation may be due to the fact that the incidence of upper respiratory tract infections is higher in our region during these months. Dysautonomia is common in severe cases of GBS and manifests as wide fluctuations of blood pressure with orthostatic hypotension, resting tachycardia and even cardiac arrhythmias. Usually ten to twenty percent of patients can develop dysautonomia. This percentage can increase to seventy five in severe cases. The afferent limb of cardiovascular regulation contains more myelinated fibers than the sympathetic and parasympathetic efferences, which determine the common classification of dysautonomia. Catecholamine levels are increased which leads to hypertension, tachycardia, ECG-changes and hyperglycemia. Norepinephrine sensitizes left ventricular stretch receptors. They induce cardiovascular depression and neurocardiogenic syncope. Many studies have found that mortality and functional outcomes are worse in those GBS patients who have dysautonomia. Therefore a close monitoring is needed in such cases and proper treatment may be provided. IVIG is preferred in presence of dysautonomia as hypotension is a common phenomenon during plasmapheresis. In our study only five patients had dysautonomia. This low incidence may have been due to less severity of GBS in our group of studied patients. The most life-threatening complication of GBS is respiratory failure due to severe weakness of the respiratory muscles. About ten to thirty percent of GBS patients may require mechanical ventilation. GBS has been reported to be the commonest peripheral neuropathy causing respiratory failure. Endotracheal intubation is more difficult in patients with GBS who have dysautonomia as sudden arrhythmia or hypotension can happen during airway manipulation. In our study only four patients needed ventilator support for respiratory failure. Fortunately none of those developed any complication during intubation. Plasmapheresis, or plasma exchange (PE) and Intravenous immunoglobulin (IVIG) are the two treatment options available for GBS. Plasma exchange nonselectively removes immunoglobulins, complement, and cytokines, all of which may play a role in the pathogenesis of GBS. PE is typically administered as 4 to 6 total plasma volumes exchanged over a span of 10 to 14 days. Alternate day PE reduces the risk of coagulopathy by permitting hepatic synthesis to regenerate serum clotting factors. It allows time for antibodies initially located outside of the plasma compartment to redistribute back into the serum, where they can be removed during subsequent exchange .The exact mechanism by which IVIG exerts its beneficial effect in GBS is not firmly established, but it may act by neutralizing autoantibodies or cytokines, saturate macrophage Fc receptors, or inhibit complement activation. In our study we found that only nine patients received disease modifying treatment. Five patients received IVIG and Plasmapheresis was done in four patients. That means only around ten percent of total GBS cases had received specific treatment. Though there are no big studies to show the exact number of GBS patients who receive IVIG/PE in other regions but we think that the number in our study is less. The reason for this could be the high cost of IVIG which cannot be afforded by all and the fact that most of the people in our region usually do not have any kind of insurance. Another reason could be that we don't have too many severe cases of GBS here.

CONCLUSION

GBS shows a lot of variation from country to country as well as in different states of same country as regards the demographic profile. Therefore it is important to know these features so that adequate measures can be taken to prevent GBS and keep the healthcare facilities ready to manage the cases during the peak season. In addition females in postpartum period being at a higher risk of GBS need to be monitored so that an early diagnosis and management can be done.

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