

Guillain-Barre' Syndrome: Clinical Profile

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ABSTRACT

Guillain-Barré Syndrome (GBS) is an acute, immune-mediated polyradiculoneuropathy and an important cause of acute flaccid paralysis (AFP) worldwide. Respiratory insufficiency requiring ventilator occurs in 30% of patients that prolong the hospital stay, leading to morbidity and mortality. There have been relatively few studies of Guillain-Barresyndrome in adults from North India.

Objective : To evaluate clinical profile, epidemiological, laboratory and electrodiagnostic feature of patients with Guillain Barre Syndrome in adults and use of intravenous immunoglobulin (IVIg) in addition to supportive care.

Materials and Methods : This was a prospective study of 50 patients with GBS admitted to our medicine department in rural tertiary care institution Dr.RPGMC Tanda. We studied the epidemiological, clinical, electrophysiological features and their outcome.

Result: In our study 50 cases were taken, motor weakness was the most common presenting feature. Antecedent illness was found in 48% of cases in the preceding two weeks, which included nonspecific illness, acute respiratory infection, diarrhea, and viral infection like chickenpox. At onset, sensory symptoms (pain and paresthesia) were noted in 16% of the cases and limb weakness in 77%. On admission, a majority (64%) was in Hughes neurological disability grading stage III-IV' all had limb weakness at the peak deficit; autonomic disturbance was seen in 35.8%, and bulbar palsy in 6%. Duration of illness was less than three weeks in 60% of cases. The one patient had undergone respiratory distress and kept on ventilator for 23 days and survived.

Conclusions: Male preponderance and motor weakness was the most common presenting illness and a majority achieved full recovery in our study. Although IVIg may be useful in the treatment of GBS, the key issue is excellent intensive care unit management.

Key words: Guillain-Barré syndrome–intravenous immunoglobulin–hughes grade–supportive care

1 INTRODUCTION:

Guillain-Barré syndrome (GBS) is the most common non-polio acute flaccid paralytic illness affecting people in the era of global eradication of poliomyelitis. GBS is an acute, monophasic, symmetrically progressive, peripheral ascending demyelinating polyneuropathy characterized by rapidly evolving symmetrical limb weakness, areflexia, absent or mild sensory signs, and variable autonomic disturbances.

Although most patients have a favorable outcome, mortality is usually related to systemic problems or complications of hospitalization, rather than the actual disease [1, 2] GBS is the major cause of acute neuromuscular paralysis, with an annual incidence of 1.3-2 per 100,000 worldwide [3, 4] Electrophysiologically most patients suffering with GBS have AIDP and motor axonal degeneration, minimal cellular inflammation, which is termed as 'acute motor axonal neuropathy' [5] Approximately two-thirds of all GBS cases are preceded by an infection such as mild respiratory infection or diarrhea [6] GBS is an acute inflamma-

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tory disorder of the peripheral nervous system thought to be due to autoimmunity for which immunotherapy is usually prescribed [7]. The clinical criteria proposed by Asbury and Cornblath are generally accepted as the guideline for diagnosing GBS. Affected adult patients usually recover in longer time than children, while the mortality rate was reported at 3-5%. [8, 9] We performed a prospective study on the natural history with GBS to study their clinical profile using intravenous immunoglobulin (IVIg) in addition to supportive care.

2 MATERIALS AND METHODS:

In this study of 50 adults above the age of 18 years with GBS admitted to the four units in the rural tertiary care institution Dr.RPGMC Kangra at Tanda H.P from March 2017 to July 2019. where 50 consecutive GBS patients, underwent detailed clinical and electrophysiological assessment. Institutional Ethics Committee approved the study protocol and written informed consent was taken from all relatives of participant. Information regarding the relevant variables in the study was collected with the help of structured proforma comprising age, sex, antecedent infections, duration of illness, seasonal trend, clinical variant, recurrence, asymmetry, CSF analysis, electrophysiological variant, Hughes disability score, treatment. Asbury and Cornblath's clinical diagnostic criteria for GBS were used for clinical diagnosis [10]. Patients with equivocal diagnosis or inadequate clinical details or laboratory investigations were excluded. The disability staging was done according to the Hughes neurological disability grading 7-point scale that was subsequently adapted by the Plasma Exchange/Sandoglobulin Guillain-Barré Syndrome Trial Group, 1997 (modifications in italics). [11, 12] Medical Research Council (MRC) sum score was used for valuing the muscle strength from 0 to 5. Classification of patients as axonal or demyelinating subtype was based on electro diagnostic criteria of Hadden et al. 9 Data obtained in the study were subjected to statistical analysis with Statistical Package for Social Sciences (SPSS) version 18. Categorical variables were summarized as counts (percentage). 32 patients were treated with IVIg in a dose of 0.4g/kg body weight over 5 days. 10 were given Intravenous Methylprednisolone along with adjunct intervention to conventional supportive and respiratory care. Rest were managed symptomatically.

3 RESULTS

A total of 50 patients were studied. All patients were hospitalized and the average duration of hospital stay was 17.57 days. Age and Sex distribution: 35 patients (70%) were males and 15(30%) were females. The age of patients ranged from 18 to 70 years (Mean age 35.72 years) with the maximum number (48%) of patients in between 40 to 50 yrs. age group & in between 30 to 40 (40%) yrs. Figure 1. Most number of cases were seen in the months of April to June. Antecedent illness: Twenty four (48%) patients had some

antecedent event prior to the development of GBS Figure 2. The most common antecedent illness was upper respiratory tract infection and diarrhea. At onset, sensory symptoms (pain and paresthesia) were noted in 8 cases (16%), and limb weakness was seen in 39 cases (76.98%). On the day of admission, 17 cases were in stage IV of illness, 32 patients in stage III, and 1 patients in stage V. Clinical features at the peak deficit were analyzed. All the 39 cases had limb weakness, Other features like bulbar palsy were present in 3 cases (6%). Autonomic disturbance was noted in 14 cases (28%). We also observed constipation and sinus bradycardia. Apart from these, bladder symptoms were noted in 3 cases (6%), transient hypotension in 5 (10%). Table 2]. In the electrophysiological studies, 20(40%) cases showed the presence of acute inflammatory demyelinating polyneuropathy (AIDP), Axonal involvement in 8 (16%) cases and mixed in 22 (44%) patients Figures 3 and 4. In the cerebrospinal fluid (CSF) study, 41 cases (38.85%) showed albumin-cytological dissociation. one case showed low CSF protein and polyneuropathy; the case was later confirmed by nerve conduction study. Researchers noted varied results on the benefit of IVIg in shortening the course of GBS, though IVIg is easily administered and well tolerated and are currently the first-line immunotherapy in GBS. 32 (64%) cases received IVIg and improvement is seen in symptom. Out of 50 patients, 32 (64%) received Intravenous Immunoglobulin (IVIg). 10 (2.7%) received I/V Prednisolone and 8(16%) did not receive any treatment either due to minimal weakness or in recovery phase.. Outcome was assessed at discharge, and at 1, 3, 6, and 12 months. 32 (64%) cases were able to walk with or without support at 3 months, and complete recovery at 6 months 34(80%), 1 case (2%) was bed bound.

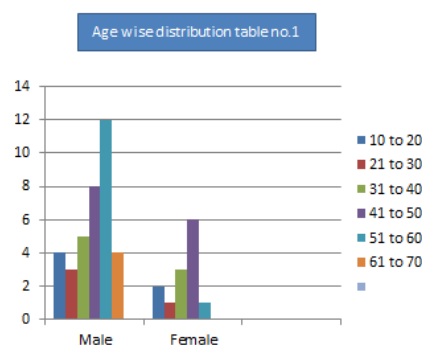


Figure 1.

4 DISCUSSION:

Fifty patients were included in this prospective study. The maximum number of patients was in between 40 to 50 years age group (28%). Kaplan et al [13] reviewed 2575 cases and

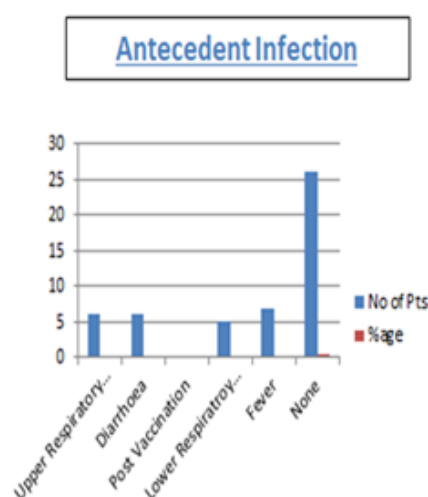


Figure 2.

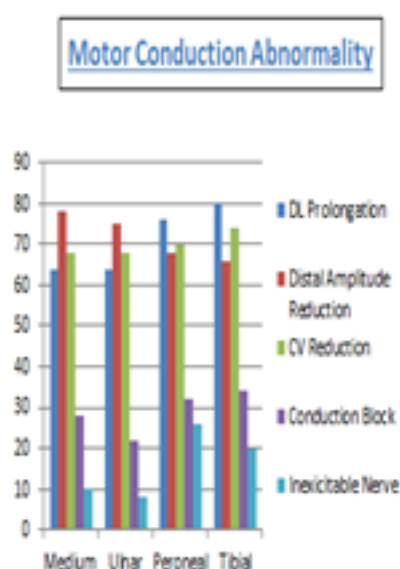


Figure 3.

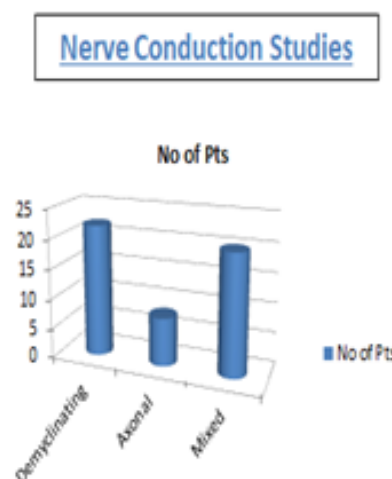


Figure 4.



found the peak incidence to be between 50 and 74 years of age with lesser peak between 15 and 35 years. Similarly Peter C. Dowling [14] also reported two peaks. There is a male preponderance in our study which is in conformity with the report by Robert M. et al. [15]. However, Peter C. Dowling's [14] study of 176 patients showed an equal incidence in males and females. No seasonal variation in incidence of GBS could be inferred from this study in conformity with the majority of studies in literature [15]. However, a few studies have noted a seasonal clustering of cases. Kaur et al [16] reported an increased incidence in summer and autumn. Peter C. Dowling [14] also noted an increase in summer. Twenty-four (48%) of our patients had a definite antecedent event prior to onset of illness. Winer et al [13] reported that over half of GBS patients experience symptoms of viral respiratory or gastrointestinal infections. Ropper et al also reported a high incidence of 73%. In contrast, a study by Kaur et al [16] showed a lower incidence of 32%. Zahahirul Islam et al showed 69% had antecedent illness of which 37% of cases were preceded by diarrhoea [17]. The interval between prodromal illness and onset of GBS is most frequently from 1-3 weeks. Occasionally it is as long as 6 weeks. Kaur et al [16] reported a mean interval of 9.2 days. In our study, there is a mean interval of 8.06 (± 4.21) days between the prodromal and the onset of GBS. The most common antecedent illness was upper respiratory tract infection (18%) while diarrheal illness and lower respiratory tract infection was seen in five patients (10%). Ascending paralysis was noted in 70% (35 patients) and descending paralysis in 4% (2 patients), while 26% (13 patients) had simultaneous involvement of all four limbs. According to description by Winer et al [13] that muscle weakness usually

starts in legs and ascends to arms in most cases. A meta-analysis of large series by Allan H. Ropper [18] showed ascending paralysis in 60%, descending paralysis in 20% and involvement of all four limbs simultaneously in 20% cases.

All patients had involvement of the legs and involvement of limbs was symmetric in all cases. None of the patients had involvement of hands alone, which is in conformity to the observation of Winer et al [13] who said that the arms are not affected in isolation. One patient also had ataxia and involvement of 3rd, 4th and 6th cranial nerves. They were diagnosed to have Miller Fisher Variant of GBS. Overall, about 50% of patients with GBS reach maximal weakness by 1 week, 70% by 2 weeks, and 80% by 3 weeks in the course of illness. 56% of our patients had cranial nerve dysfunction. 20% of patients had involvement of multiple cranial nerves. This is in conformity with the 50% incidence reported by Winer et al [13] and 60% in Allan H. Ropper's meta-analysis. Kaur et al [16] reported an incidence of 41% in her study from North India. Facial Nerve was the most commonly involved 22 patients in our series in concordance with most series. IX and X cranial nerves were involved in only 9 patients in contrast to the reported incidence of 50% in Allan H. Ropper's meta-analysis. 3, 4, 6 cranial nerves involved in 2 patients. In this study autonomic dysfunction occurred in 28% of patients. Cardiac arrhythmia occurred in 6% of cases, postural hypotension in 10% of cases, Fluctuating Blood pressure was noted in 8% of patients. Transient sphincteric dysfunction in the form of urinary retention and hesitancy was seen in three (6%) patients. Allan H. Ropper's meta analysis reported 15% incidence of transient bladder disturbances in GBS patients NK Singh et al [12] observed sphincter disturbance in 20% of patients.

CSF protein was raised above 50mg% in 41 (82%) patients. Winer et al [13, 19] reported raised CSF protein in 80% patients while 90% was reported in Allan H. Ropper's [18] meta-analysis. The lower number of patients with raised CSF protein in this study was probably because all CSF studies were done between 1 and 2 weeks from onset and not repeated thereafter. It is possible that there may have been a rise in CSF protein later in the course of illness, which was not recorded. Furthermore, it has been noted in some studies that CSF protein may not rise throughout the course of illness in some patients with GBS. Gupta RC17 [19] reports such patients did not show rise in CSF protein even at 6 weeks. CSF pleocytosis was seen in three patients. Electrophysiological studies were conducted in all patients and 20 of them showed demyelinating pattern, 8 of them showed axonal pattern, 22 patients mixed pattern. Patients having mixed and axonal pattern showed poor prognosis compared to patients having demyelinate pattern.

5 CONCLUSION:

Males were predominantly affected. Motor weakness was the most common presenting illness. Female preponderance was observed in demyelinating form and male preponderance in

axonal form of GBS. AIDP patients were older, while axonal form were younger. Seasonal occurrence predominantly in rainy season was noted. Axonal (AMAN+ AMSAN) form dominated than demyelinating form (AIDP). GBS occurs in all age groups with a greater incidence in the older age group above 40 years. However age did not have any correlation with prognosis. GBS affects both sexes; however males were affected more than females in the ratio of 3.6:1.4 in this study. 48% of patients reported a definite antecedent event prior to onset of GBS. Onset of GBS is heralded by both motor and sensory symptoms. Ascending type of paralysis was most commonly seen in our study. Peak flow test, peripheral saturation, single breath count, and bulbar weakness may be a predictor of respiratory failure. There was a significant association of Low MRC sum score with respiratory failure. Early diagnosis of respiratory failure and prompt intervention improves patient outcome. Further large sample studies are required to assess respiratory failure and subdivision of Hughes grade 5.

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