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To Study The Indicators For Severe Course of Guillain-Barre Syndrome.

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ABSTRACT

Background: Guillain-Barré Syndrome (GBS) is an acute ascending flaccid paralysis, often preceded by a mild bacterial or viral infection. Aim: To study predictive indicator for the severe course & mortality in patients with GBS. **Methods:** Prospective follow up study of 50 EMG NCV proved GBS patients admitted in paediatric department of civil hospital during 1 Nov 2015 to 31 Oct 2016. Details of all patients were noted in prestructured proforma during their course of illness and follow up. **Results:** Mean age of patient in study was 5.14±2.5 years with male to female ratio 1:1.38. Preceding infection or vaccination was detected in 33(66%) cases out of which respiratory infection (45.4%) was most common followed by gastrointestinal infection (30.3%) and following Vaccination (3.1%) Most common nerve conduction abnormality noted was acute inflammatory demyelinating polyradiculoneuropathy(AIDP)28 patients(56%), followed by acute motor axonal neuropathy(AMAN)20 patients(40%), and acute motor-sensory axonal neuropathy(AMSAN)2 patients(4%).GI infection was most commonly associated with AMAN(60%) findings with longer duration of recovery (walking with aid mean no. of days: 86±34.3 days & without aid: 139±46.4 days) while respiratory infection was associated with AIDP(53.3%) with relatively shorter duration of recovery(walking with aid mean: 34±19.3 days & without aid: 62±27.6 days). Mortality was higher in patients with GI symptoms (40%) & AMAN (25%) as compare to respiratory infection (20%) & AIDP(17.8%). Ventilatory support required more in patients who had speech impairment(66.6%) as compare to autonomic disturbance(61.5%).Mortality was higher in patient who had speech impairment (5/9,55.5%) as compare to autonomic disturbance(6/13,46%). **Conclusion:** Preceding history of GI infection, AMAN and speech impairment at the time of presentation or during the course of disease are predictive indicators for severe course of GBS in children.

Keywords: EMG-NCV, Guillain-Barré syndrome, Preceding infection, Speech impairment.

INTRODUCTION

Guillain-Barré syndrome (GBS) is an autoimmune disorder often considered post infectious polyneuropathy involving mainly motor but also sensory and sometimes autonomic nerves. It is characterized by rapidly progressing symmetrical muscle weakness and loss of deep-tendon reflexes (DTR).It is now the most common cause of acute flaccid paralysis worldwide since the near-demise of polio.[1,2] There are four main subtypes of GBS according to clinical and pathological features: Acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor-sensory axonal neuropathy (AMSAN), and Miller Fisher syndrome (MFS).[3] India is far behind in GBS research as compared to other countries as there might be a low case report of GBS in Indian population. Every study and different region within the same country has a different age of presentation, clinical features and electrophysiological types of GBS. By understanding the risk factors & use of these factors makes it possible to predict the prognosis of GBS patients, and to identify patients with poor prognosis in the early phase of the disease and provide these patients with intensive treatment.

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MATERIALS AND METHODS

Prospective follow up study of EMG NCV proved GBS patients admitted in paediatric department of civil hospital, Ahmedabad during 1 Nov 2015 to 31 Oct 2016. Patient of GBS with age 1 month to 12 years and with EMG NCV finding were included in study. Patients were excluded if they had any one of the following: age more than 12 years, case of chronic inflammatory demyelinating polyradiculopathy or any other cause of acute flaccid paralysis and clinical GBS without EMG-NCV findings .A written informed consent was taken from

all the patients. For the present study, the clinical details for each patient were recorded in prestructured proforma, including age, gender, antecedent infections, time from onset to admission, clinical severity, muscle weakness, sensory disturbance and reflexes in arms and legs, cranial nerve deficits, autonomic disturbance and pain, as well as treatment modality and complications during hospitalization were collected. Clinical severity of patient was assessed by Hughes Function grading scale and muscle weakness by medical research council sum score.[4,5] Nerve conduction studies were done to determine GBS subtypes and clinical correlation with electrophysiological features was done. EMG NCV was recorded with 2/4 Channel Computerized EMG/NCV/EP System available in hospital. Treatment like intravenous immunoglobulin (IVIG) & plasmapheresis were given according to its availability and weight of the patient. Ventilator care and supportive treatment were provided as an when required. Patients were followed up for muscle weakness till full recovery achieved or upto 1 year after discharge, whichever was earlier.

RESULTS

During the study period 50 patients with male to female ratio 1:1.38 were enrolled in this study with Mean age of presentation was 5.14 ± 2.5 years. History of preceding infection or vaccination was detected in 33(66%) patients out of which respiratory infection (45.4%) was most common followed by gastrointestinal infection (30.3%). History of recent vaccination of measles was present in 1 patient (3.1%).At the time of admission, all patients had lower limb muscle weakness and absent DTR. There was paresthesia and pain in 38 (76%), speech impairment in 2 (4%), autonomic disturbance in 3 (6%) and respiratory failure in 5 patients (10%) at the time of admission. Detailed clinical course and various morbidities developed during the course of illness are described in [Table 1].

Table 1: Clinical course and various morbidities during hospitalization.

Signs	No. of patients (n=50)	Patients require ventilatory support	Mortality (n=10)
Quadriplegia	28	11	8(28.5%)
Paresthesia and pain	38	10	9(23.6%)
Bulbar palsy	9	6	5(55.5%)
Other cranial palsy	4	1	1(25%)
Respiratory failure	14	11	4(28.5%)
Autonomic disturbances	13	8	6(46%)

Clinically, most common variety was ascending type (86%) followed by descending type (12%) and Miller-Fisher variant (2%). [Figure 1]

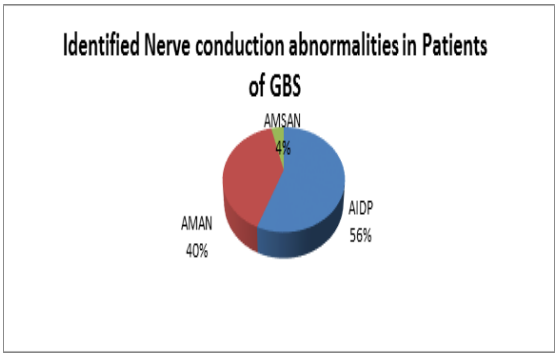

Figure 1: Identified Nerve conduction abnormality in patients with GBS.

27(54%) patient were treated with IV IG, 15(30%) were undergone plasmapheresis and 8 (16%) were treated with both. Plasmapheresis followed by IV IG was given in patients who had rapid progression of disease, no improvement or deterioration after 5 cycles of plasmapheresis and patient had autonomic disturbance leading to hypotension. During the course of illness, 40(80%) patients were discharged with mean duration of hospital stay in discharge patients 9.76 ± 6.79 days and 10(20%) were expired due to complications. Details of the antecedent events, types of GBS and morbidity described in [Table 2].

Table 2: Correlation of Antecedent illnesses with GBS subtypes & mortality.

Antecedent illnesses	Number of patiebts (n =50)	AIDP (n=28)	Aman (n=20)	AMSAN (n=2)	Mortality (n=10)
No h/o antecedent illnesses	17	9	7	1	2
Bodyache	4	3	1	0	0
URTI	15	8	6	1	3
Diarrhea	10	4	6	0	4
Recent Vaccination	1	1	0	0	0
Fever	3	2	1	0	1
Total	50	28	20	2	10

Out of 50 patients, 10 patients were expired during hospital stay. Most common cause of mortality was autonomic disturbance in 6 patients followed by bulbar palsy in 5 patients and respiratory failure in 4 patients. Mortality was higher in patients who presented with speech impairment(50%) & autonomic disturbance(33%) at the time admission.

Table 3: Duration between onset of illness & hospitalization and correlation with mortality.

Duration between onset of illness & Hospitalization	No. of Patients	Mortality
<24 hours	15	4(26.6%)
24-48 hours	9	2(22.2%)
48-72 hours	16	3(18.7%)
4-7 days	7	1(14.2%)
>7 days	2	0

Out of Total 40 patients discharged, 38 patients were fully recovered in 1year follow up but 2 patients were having residual weakness at the 1 year follow up. Recovery in patients with GI Infection, walking with aid was 72±28.4 days & without aid were 130±42.6 days. Recovery in patient with respiratory infection with aid was 38±13.8 days & without aid were 70±26.6 days. Patient with AMAN variety had mean duration of walking with aid were 86±34.3 days and without aid were 139±46.4 days while patient with AIDP variety had mean duration of walking with aid were : 34±19.3 days & without aid: 62±27.6 days. Overall, mean duration of recovery in 40 patients by walking with aid was 57.5± 22.4 days and mean days of recovery without aid was 100.25± 36.8 days.

DISCUSSION

French physician Jean Octave Landry first described the disorder in 1859. Later in 1916, George Guillain, Jean Barre and Andre Strohl diagnosed in two soldier and described the key diagnostic abnormality-albuminocytological dissociation in CSF in Guillian Barre syndrome.[6] GBS has a worldwide distribution with an annual incidence of approximately 1.2-8.6 cases per 100,000 people.[7] The incidence of GBS in India is 0.4 to 4.0 per 100,000 per year.[8]

The mean age of presentation of GBS in this study is 5.14±2.5 years which is lower than that of Meena et al (8 years) and Sarkar et al (6.5 years).[9,10] The lower mean age of presentation due to variation in infectious agent and its disease epidemiology according to region. There was history of antecedent illness in majority of patients ie. 33 (66%) which was comparable to Sarkar et al (58.3%) but higher than meena et al(48.8%).[9,10] Again the incidence of URTI in the study (30%) is comparable to that of Sarkar et al (25.3%) and Meena et al (25.6%).The incidence of diarrhea in the presence study was 20%. However, incidences of diarrhea in Sarkar et al was 12% which is lower than the present study and 30.2% in Meena et al which was higher.[9,10] The present study was done in western region of country while sarker et al and meena et al were done in east & south region respectively. The incidence of paresthesia and pain in this study was 68%which was lower than Sarkar et al (89%) & higher than Meena et al (41.8%).[9,10] Present study shows respiratory involvement in 10% of the patients at the time of admission. Respiratory compromise was noted in 28.1% of patients of Sarkar et al while 9.3% only in Meena et al at the time of admission.[5,10]

This depends on immunology, socioeconomic and geographical variation.Bulbar Palsy in present study was 18% which is reported higher in Sarkar et al (51.5%) and Meena et al (32.5%).[9,10] This may be due to early diagnosis & intervention leading to arrest of progression of weakness. 26% of patient this study developed autonomic disturbance whereas Meena et al reported autonomic involvement in 13.9% of patients and Sarkar et al has reported in 35.2% patients.[9,10] This wide variation of autonomic disturbances may be explained by different immunology and infectious profiles existing at specific geographical area and requires further study.

In present study, AIDP contributes to 56% of the patients while AMAN & AMSAN is diagnosed in 40% and 4% respectively, which is almost similar in Meena et al.[9] GI infection was most commonly associated with AMAN (60%) whereas respiratory infection was associated with AIDP (53.3%), this results were also supported by a study in northern India.[11,12]

Pediatric GBS patients require respiratory support in 15-20% of cases.[3] Similar to literature, 11 of our patients (22%) had to be put on respiratory support. 6 of them had speech impairment. Bulbar involvement-related palate dysfunction can lead to speech impairment and severe respiratory distress. It has been demonstrated that the need for respiratory support can be foreseen in GBS if there is bulbar involvement.[13] This confirms that clinical findings are very important in predicting the course of the disease. Similarly, in western studies cranial nerve involvement was more common in children who required mechanical ventilation & progression to mechanical ventilation was highly likely to occur in those patients with rapid disease progression, bulbar dysfunction, bilateral facial weakness, or dysautonomia.[13,14]

Patients with AMAN findings which was associated with GI infection had longer duration of recovery (walking with aid mean no. of days: 86±34.3 days & without aid: 139±46.4 days) as compare to AIDP which was associated with respiratory infection (walking with aid mean: 34±19.3 days & without aid: 62±27.6 days). The fact that those of our patients with history of acute gastroenteritis took significantly longer to recover and begin walking aided/unaided can be attributed to them having the AMAN subtype.[15]

Mortality was higher in patients with GI symptoms (40%) &AMAN (25%) as compare to respiratory infection (20%) &AIDP (17.8%).Mortality was higher in patient who had speech impairment associated with bulbar palsy (5/9,55.5%) as compare to autonomic disturbance (6/13,46%). Mortality was higher in patients who had rapid progression of weakness within 24 hours involving all four limbs, speech impairment and with autonomic disturbance at the time of admission.

The main limitation of our study was its lack of control group & long term follow up; involvement of various confounding factors and multifactorial risk factors which cannot be studied individually.

CONCLUSION

Preceding history of GI infection, AMAN, rapid progression and speech impairment at the time of presentation or during the course of disease are predictive indicators for severe course of GBS in children, which require stringent monitoring, and aggressive management of disease in order to reduce morbidity and mortality.

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