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STUDY OF CLINICAL PROFILE OF GUILLIAN BARRE' SYNDROME WITH SPECIAL REFERENCE TO NERVE CONDUCTION STUDY

Paediatrics

KEYWORDS: Abbreviations

GBS– Guillian-Barre syndrome CNS- Central Nervous SystemEMG-NCV-electromyogram and nerve conduction studies IVIG- immunoglobulins AIDP-acute inflammatory demyelinating polyneuropathy AMAN-acute motor axonal neuropathy AMSAN-acute motor sensory axonal neuropathy

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ABSTRACT

Aims and objectives: To study incidence, various clinical presentations, and patterns in electrophysiological variants of Guillian-Barre syndrome. And To compare outcome of various types of GBS. **Setting & Design:** The prospective study was conducted in the Pediatric department of tertiary care hospital and medical college of western India, studied from 1st September, 2013 till 30th may 2016 . **Methods & Material:** All clinically diagnosed cases of GUILLIAN BARRE' syndrome as per Asbury's criteria between 1-12 years were included .EMG-NCV was done to assess the variety of GBS. In age below 3 years or a child weighing less than 12 kg was offered IVIG while in others plasma pheresis was offered .Results were analyzed. **Results:** GBS constituted 0.50% of all admitted patients and 9.5% of all neurological patients.In our hospital. The mean age of presentation is 3.8 years of age in this study with the youngest being 13 months. Overall male to female sex ratio is 1.49:1 in present study which shows male preponderance. In our study, limb weakness was the most common complaint for admission found in all 137 patients which was most commonly affecting lower limbs (57.6%) followed by all four limbs in 40.22%. In present study, classical variant was most common found in 92.3% of the patients make shows ascending type of paralysis. Based on EMG NCV results, AMAN is the most common electrophysiological type of GBS reported for 54.8% followed by AIDP for 38.8%. In present study, 76 patient had received intravenous immunoglobulins (IVIG) within 72 hours of onset of symptoms which accounts for 55.5% with survival of 53 patients (70.6%).IVIG is the most commonly used modality of treatment in present study, due to younger age group were plasmapharesis was not possible. Plasmapheresis has been performed in only 27 patients (19.5%) which survival of 77.6% of the patients

Introduction

GBS has a worldwide distribution with an annual incidence of approximately 1.2-8.6 cases per 100,000 people. Both genders are at similar risk (but there is a slight male predominance). All ages are affected, although the distribution is bimodal. GBS is an autoimmune disease that affects peripheral nerves. The immune response is misdirected to , and the immune system turns against peripheral nerves, most often the myelin sheath.In GBS, about two-thirds to three-quarters of the people affected are able to identify an illness or event that occurred a week or two prior to the development of weakness. Most commonly, this illness is an infection(influenza like illness, campylobacter jejuni,viral illness like Cytomegalovirus, Mycoplasma pneumonia, Epstein Barr virus and HIV),GBS can also occur after some vaccinations (tetanus toxoid containing vaccines, measles vaccine, hepatitis B vaccine)and following surgery or other trauma. The neurologic symptoms appear an average of 2 weeks after the triggering event, although they may appear as early as 1 week and as late as 4 weeks. One consistent observation is that the shorter the interval between the antecedent event and the onset of neurologic symptoms, the more severe the GBS .

Clinical Features include Muscle weakness which is ascending and symmetrical type of paralysis affecting lower limb paralysis and then affects upper part of body. This is often described as a "rubbery" feeling. Cranial Nerve Involvement in form of facial weakness , of the ninth and tenth cranial nerves. leading to difficulty in swallowing and handling saliva. In extremely severe cases,there may be loss of all voluntary muscle movement and the person will not be able to communicate. This is sometimes called the locked-in syndrome. Sensory symptoms occur in 50 to 70 percent of GBS patients. People may also complain of numbness or loss of sensation. In about 30 percent of GBS patients, there is painful muscle cramping between the shoulder blades or in the lower back or thighs, and the muscles may be tender.Some people experience a sensation of tiny insects crawling over their skin, a condition called formications.Perhaps a more common manifestation of the loss of sensation is ataxia, an imbalance that can occur in any case of GBS. This is particularly prominent in Miller Fisher syndrome. Loss of proprioception accounts for the ataxia of Miller Fisher syndrome.Areflexia and hyporeflexia are major findings in GBS. The finding of normal reflexes will rule out GBS.Autonomic Disturbance like Urinary retention, Constipation or paralytic ileus, Syncope with low blood pressure, Pounding headache with elevated blood pressure, Palpitations, Confusion or seizures caused by low blood salt (sodium) level. Increased sweating, Bluish discoloration and cold temperature due to vasomotor instability. Guillain- Barre syndrome (GBS) is generally diagnosed on clinical grounds.

Basic laboratory studies, such as complete blood counts (CBCs) is inconclusive. special investigations like Electromyography (EMG) and nerve conduction studies (NCS) with typical changes documented in late phase only (3-4 weeks). Lumbar puncture for cerebrospinal fluid (CSF) studies is recommended during second week of GBS, characteristic findings on CSF analysis include albuminocytologic dissociation. Serum autoantibodies are not measured routinely in the workup of GBS, but results may be helpful in patients with a questionable diagnosis or a variant of GBS. The two major aspects in the treatment of GBS are Specific Therapy Immunomodulation in form of Plasma exchange, Intravenous immunoglobulin, Steroid therapy, Combined therapy; and supportive treatment. Prognosis of the disease is good with complete recovery if intervention done at the earliest. Clinical feature predictive of poor outcome with sequelae are cranial nerve involvement, intubations and maximum disability at the time of presentation.

Materials And Methods: The prospective study was conducted in the Pediatric department of tertiary care hospital and medical college of western India. All clinically diagnosed cases of GUILLIAN BARRE' syndrome as per Asbury's criteria between 1-12 years Total indoor cases were studied from 1st September, 2013 till 30th may 2016.Patient's name, age, sex and indoor registration number, duration of hospital staywere noted. They were inquired in detail about their presenting illness and any preceding illness. History of immunization was taken in detail in all patients. In general examination vital signs were monitored. Patients were monitored for autonomic disturbances.Thorough neurological examination as well as other systemic examination was done.Widely adapted disability

grading scale of Hughes and colleagues was used in defining the functional motor deficits of patients.GBS was diagnosed clinically through history and clinical examination. Relevant investigations were done.According to age, weight, economic status and general condition of patients IVIG or plasma pheresis was given.In age below 3 years or a child weighing less than 12 kg was offered IVIG while in others plasma pheresis was offered.All data are analyzed with the help of new GraphPad demo version. EMG NCV was recorded with 2/4 Channel computerized EMG/NCVsystem available in physiotherapy department. Patients were reassessed clinically during admission, on discharge and periodically or 2 months whichever is earlier.

RESULTS

During the study period, total indoor pediatric cases were 27346, GBS constituted 0.50% of all admitted patients and 9.5% of all neurological patients.In our hospital, only children up to 12 years of age are examined in pediatric department. In this study, 45% of the patients were from 1-5 years of age with maze to female sex ratio of 1.52:1 ; in 6-10 years of age group, sex ratio is 1.36:1 with total number of patients of 59(43%).Sex ratio is higher in older age group of 11-12 year (2:1) with total number of patients of 15 (10%).The mean age of presentation is 3.8 years of age in this study with the youngest being 13 months. Overall male to female sex ratio is 1.49:1 in present study which shows male preponderance. In present study, incidence of GBS is almost constant throughout the year, as 37% of cases were noted in summer and 39% in rainy seasons. If however, shows a decrease in the winter months (24%) as compare to the rest of the year.

In our study, limb weakness was the most common complaint for admission found in all 137 patients which was most commonly affecting lower limbs (57.6%) followed by all four limbs in 40.22%. 2.1% patients presented with upper limb weakness at onset. Bulbar palsy was seen in 22.8%. Sensory symptoms like pain and paresthesia were present in 67.4% patients. Significant bowel and bladder involvement was present in 27.2% patients with bladder involvement more commonly. 21.7% patients were presented with complaint or respiratory distress..

In present study, autonomic disturbances are observed in 25% patients. 73.9% had tachycardia, while 13.1% had intermittent bradycardia and 8.7% patients developed hypotension while 4.3% had hypertension.

In present study, classical variant was most common found in 92.3% of the patients make shows ascending type of paralysis.Three patients (2.2%) were presented with descending paralysis.Three patients (2.2%) were presented with predominantly sensory symptoms. Three patients were diagnosed as Miller fisher variety. One patient also presented with asymmetrical paralysis (right >left)
In present study EMG NCV was done in 110 patients. It was not possible in all patients due to infrastructure limitation and poor general condition of some patients. Based on EMG NCV results, AMAN is the most common electrophysiological type of GBS reported for 54.8% followed by AIDP for 38.8%. 3.2% patients were diagnosed as AMSAN by electrophysiological study.3.2% patients remained unclassified as their EMG NCV test were normal.

In present study, 76 patient had received intravenous immunoglobulins (IVIG) within 72 hours of onset of symptoms which accounts for 55.5% with survival of 53 patients (70.6%).IVIG is the most commonly used modality of treatment in present study, due to younger age group were plasmapharesis was not possible. Plasmapheresis has been performed in only 27 patients (19.5%) which survival of 77.6% of the patients. Three patients who went through plasmapheresis were not improved and looking at their prognosis. IVIG was also given, all got discharged.31 patients were only on supportive treatment and no immune modulation was offered because of static nature of disease observed in them. All of them discharged with little but improving weakness.

Table 1 Age and Sex distribution

Age group	Male (n=82)	Female (n=55)	Total (n=137)	Sex ratio	p value
1-5 years	38	25	63 (45%)	1.52	0.25 (NS)
6-10 years	34	25	59 (43%)	1.36	
11-12 years	10	5	15 (10%)	2	
Mean age of presentation				3.8 years	

Table 2 Presenting symptoms in patients of GBS

Presenting complaint	No. of pt (n=137)	Percentage (%)
Limb weakness	137	100
Upper limb + Lower limb weakness	55	40.2
Lower limb only	79	57.6
Upper limb only	3	2.1
Bulbar palsy	31	22.8
Paresthesia and Pain	92	67.4
Bladder/bowel involvement	37	27.2
Respiratory distress	30	21.7
Autonomic disturbances	34	25

Table 3 Electrophysiological types of GBS

Types	No. of patients (n=110)	Percentage
AIDP	43	38.8%
AMAN	60	54.8%
AMSAN	4	3.2%
Non classified	3	3.2%

Table 4 Treatment given to patients of GBS

Treatment	No. of patients (n=137)	No. of survived patientys (n=108)	Percentage of survival
IVIG	76 (55.5%)	53	70.6%
Plasmapheresis	27 (19.5%)	21	77.7%
Plasmapheresis+ IVIG	3 (2.2%)	3	100%
Only supportive treatment	31(22.8%)	31	100%
Assisted ventilation	40 (29.3%)	7	18.5%

* Mean Duration of Hospital Stay in Discharge patients – 9.76±6.59 days

Table 5 Comparison of major electrophysiological types, clinical presentation and outcome (n=58)

	AMAN (n=60) (percentage)	AIDP (n=43) (percentage)	P value
Age wise incidence			
1-5 years	28(47.1)	27(62.5)	0.40 (NS)
6-10 years	25(41.2)	14(33.3)	
11-12 years	7(11.7)	2(4.2)	
Sex distribution			
Male	35(58.8)	34(79.2)	0.10 (NS)
Female	25(41.2)	5(20.8)	
Sex ratio	1.4	3.8	
Antecedent illnesses			
URTI	5(8.8)	9(20.8)	0.22 (ns)
GI symptoms	10(17.1)	5(12.5)	
Signs and symptoms			
Motor	60(100)	43(100)	
Sensory	33(55.8)	37(87.5)	0.019(S)
Cranial nerve involvement	14(23.5)	12(29.1)	0.76(NS)
Bulbar	10(17.6)	12(29.1)	0.34(NS)
Autonomic involvement	7(11.7)	3(8.3)	1(NS)

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Respiratory distress	14(23.5)	9(20.8)	1.0(NS)
Mean Hughes grade at admission	3.2±1.0	2.6±1.1	NA
Treatment given			
IVIG	28(47.1)	25(58.3)	NA
Plasma exchange	17(29.4)	5(12.5)	
P.E.+IVIG	0(0)	3(4.1)	
Only supportive	14(23.5)	11(25)	
Assisted ventilation	10(17.6)	6(12.5)	
Outcome			
Discharged	48(79.5)	39(91.6)	0.18(NS)
Expired	9(14.7)	2(4.1)	
DAMA	3(5.8)	2(4.1)	Na
Mean duration of hospital stay in discharged patients	8.4±0.58 days	9.7±2.06 days	NA

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