

Guillain Barre Syndrome: Major Cause of Acute Flaccid Paralysis in Children and Adolescents of Nepal

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Abstract

Introduction. Guillain Barre Syndrome (GBS) is a post infectious polyneuropathy involving mainly motor but sometimes sensory and autonomic nerves. It is an acquired disease of the peripheral nerves that is characterized by rapidly progressing paralysis, areflexia and albumino-cytological dissociation in CSF.

Methodology: Prospective, descriptive, observational, hospital based study was carried out to find out the clinico-epidemiological features of GBS including existing treatment modalities and its outcome. All cases fulfilled the criteria for AFP (Acute flaccid Paralysis) surveillance was included. Cases were reviewed for full medical history and examinations. To confirm the diagnosis, necessary investigations were carried out and combined with clinical symptoms. **Results:** Thirty patients were included in the study during study period. Among them 90% were diagnosed as GBS, 7.4% patients of GBS were associated with hypokalemic paralysis, 7.4% diagnosed as transverse myelitis and 3.7% diagnosed as idiopathic neuropathy. Different types of GBS were classified as AIDP (Acute inflammatory demyelinating polyneuropathy) 62.96%, AMAN (Acute motor axonal neuropathy) - 25.52%, AMASAN (Acute motor and sensory axonal neuropathy) - 3.3% and MFS (Miller fisher's syndrome) - 6.6% according to NCV result. Male female ratio is 1.7:1.0. There was 14.8% patients had relapse within 5 year. Associated diseases were URTI, pneumonia, sore throat and diarrhea. Facial Nerve palsy was commonest cranial nerve involvement. Sixty percentage of patients presented with sensory symptoms. There was transient bowel and bladder involvement in 20% of the cases. 69.2% patients became bed ridden at the nadir. There was albumin-cytological dissociation in 80% case. Majority of patients improved with supportive treatment alone, 19.5% patient required ventilator support among them 40% died. 7.4% of cases expired during treatment. Half of the patients fully recovered within 3 months. **Conclusion:** GBS is the commonest cause of AFP, AIDP being commonest subtype in our setting. We have to improve our existing treatment facilities and extend to different centers to detect and treat GBS. Most of the patients improve with supportive treatment alone. Ventilator support indicates grave prognosis.

Key words: GBS (Guillain Barre Syndrome), AFP (Acute flaccid Paralysis), AIDP (Acute inflammatory demyelinating polyneuropathy), AMAN (Acute motor axonal neuropathy), AMASAN (Acute motor and sensory axonal neuropathy), MFS (Miller fisher's syndrome).

Introduction

Guillain Barre Syndrome (GBS) is a post infectious polyneuropathy involving mainly motor but sometimes also sensory and autonomic nerves¹. GBS is an acquired disease of the peripheral nerves that is

characterized clinically by rapidly progressing paralysis, areflexia and albumino-cytological dissociation². Earliest description of this disease is probably that of Word Rob and Oliver in 1834, but the syndrome as first described in 1859 by Jean Baptiste Octave Landry, who identified

the clinical features of the disease and termed it as 'Acute ascending paralysis. Osler reported the same disease in 1982 as febrile polyneuritis³. In 1916, Guillan, Barre and Strohl published their observation of the albumin cytological dissociation on cerebrospinal fluid examination of two shoulders who had presented with paralysis and areflexia². A comprehensive pathological account of GBS was that of Haymaker & Kernohan (1949) who stressed that edema of the peripheral nerves was the important changes in the early stage of disease³. Asbury, Arnoson and Adam (1969) established the essential lesion; it is a perivascular mononuclear inflammatory infiltrate of the root and nerves³. In 1980s and 1990s saw the development of plasma exchanges, intravenous immunoglobulin and all of which reduced the mortality and morbidity². The watershed development in the last decade, however, has been the characterization of variants of GBS and recognition of pathological association of *campylobacter jejuni* infection with the motor axonal form of GBS³.

GBS occurs in all age groups and has an annual incidence varying from 0.4 to 1.7 cases per 100000 population. The reported incidence for children under 15 years is similar to adults². It occurs in all part of the world in all seasons, affects both children and adult of both sex². In post polio era, it is the most common cause of an acute generalized paralysis². Though GBS is non-seasonal and non-epidemic, the existence of an axonal variant of GBS has been confirmed by observations on an annual seasonal epidemic that occurs in northern China in summer⁴. There is a slightly male predominance with male to female ratio of 1.5:1. while most of the cases occurs sporadically; occurrence of epidemic clusters of GBS has been described². According to the Acute flaccid Paralysis (AFP) surveillance record of Nepal there are 192, 214 and 224 cases of AFP in 2060 (2003-2004), 2061(2004-2005) & 2062 (2005-2006) respectively⁵. World Health Organization (WHO) has established standards to check the efficacy of surveillance systems in each country, polio-endemic or not, should be able to detect at least one case of AFP not caused by polio every 100,000 children under age 15. If a country can detect AFP causes corresponding to this rate, one can be reasonably certain that all polio cases among these paralyzed children have been identified⁶. Only one cases of polio was recorded in 2006, other cases of AFP neither were nor categorized properly. As post polio era GBS is the commonest causes of AFP in developed countries. Most of the cases in our setting are also assumed to be GBS.

Materials and Methods

Prospective, descriptive, observational, hospital

based study was carried out at to find out the clinico-epidemiological features of GBS in eastern part of Nepal including clinical presentations, risk factors, existing treatment modalities, prognosis of the disease in relation to its severity and outcome in the tertiary care centre. All cases fulfilling the criteria for AFP surveillance were included in study. Children enrolled in study were seen in emergency, outpatient department (OPD), Pediatric wards and pediatric intensive care unit (PICU). Cases were reviewed for full medical history, general physical and detail systemic examination, in order to confirm the diagnosis, necessary investigations were made. All children up to 15 years, coming under AFP surveillance were included. Features required for Diagnosis includes progressive weakness of both legs and arms and areflexia⁷. Clinical features supportive of diagnosis includes progression over days to a couple of weeks, relative symmetry of signs, mild sensory symptoms or signs, cranial nerves involvement (bifacial palsies) Recovery beginning 2-4 wks after progression ceases, autonomic dysfunction, and absence of fever at onset⁷. Laboratory features supportive of diagnosis includes elevated CSF protein with < 10 cells/micro liter and electro diagnostic features of nerve conduction slowing or block⁷.

Statistical Analysis

The data collected were compiled and entered in MS Excel. Statistical analysis was carried by using SPSS pc+ 11.0 versions to find out different type of correlation and statistical values. P value < 0.05 was taken as a statistically significance.

Results

Thirty patients were included in the study during 16 months of study period. Among them 27 (90%) were diagnosed as GBS, 2 (7.4%) patients of GBS were associated with hypokalemic paralysis, 2 patients (7.4%) diagnosed as transverse myelitis and 1 patient (3.7%) diagnosed as idiopathic neuropathy.

All of 27 GBS patients had undergone NCV test and classified as AIDP (Acute inflammatory demyelinating polyneuropathy)-17 patients (62.96%), AMAN (Acute motor axonal neuropathy) - 7 patients (25.52%), AMASAN (Acute motor and sensory axonal neuropathy) -1 patient (3.3%) and MFS (Miller fisher's syndrome) - 2 patients (6.6%) according to NCV result.

GBS is commonest cause of AFP, AIDP being commonest subtype in our setting. There was associated preceding illness in GBS. Majority patients presented with symmetrically ascending paralysis with gradual onset involving all limbs. Male were more affected in

present study with male female ratio as 1.7:1.0. There was no significant age distribution but slightly higher patient from pre adolescent to young adults were found. There was no seasonal variation, no significant status with immunization, but 14.8% patients had relapse within 5 year. Associated diseases were URTI, pneumonia, sore throat, diarrhea in majority of patients.

Facial Nerve palsy was commonest cranial nerve (CN) involvement, 60% of patients presented with sensory symptoms like pain, tingling and numbness. There was transient bowel and bladder involvement in 20% of the cases. All patients became areflexic during the course of illness with 69.2% becoming bed ridden at the nadir. 77.8% patients had elevated CSF protein and 88.2% patients had less than 10 CSF lymphocytes after 2 week of illness.

Nerve Conduction Velocity Study

NCV	Frequency (n = 27)	Percent (%)
AIDP	17	63.0
AMAN	7	25.9
AMASAN	1	3.7
MFS	2	7.4

63% patients had acute inflammatory demyelinating polyradiculopathy. 23 patients required only supporting

treatment including steroids and physiotherapy but five patients required ventilator support. Two patients were expired during treatment (On Ventilator due to respiratory failure). 63% of patient discharged within 1 week of admission. Hospital mortality was 7.4% and 85.18% were discharged after improvement. Out of two one patient expired after 32 days due to secondary infection with aspiration, next patient expired immediately after admission on day 1 due to autonomic instability. Hughes scale was used for monitoring of weakness, 43.47% patients became healthy at 3 month of illness; other patients were improving only 8.69% patients remain bed ridden without further deteriorations.

There was no significant correlation found between different modes of clinical presentation like duration of weakness, type of weakness. Involvement of cranial nerve was analyzed with SPSS pc+ 11.0 version and no significant correlation found. There was albumin-cytological dissociation in 80% case. Majority of patients improved with supportive treatment alone, 19.5% patient required ventilator support among them 40% died. Majority of patient discharged within 1 week of admission with some improvement. 7.4% of cases expired during treatment, at 3 month follow-up almost half patients recovered fully and remaining patients improved on follow up.

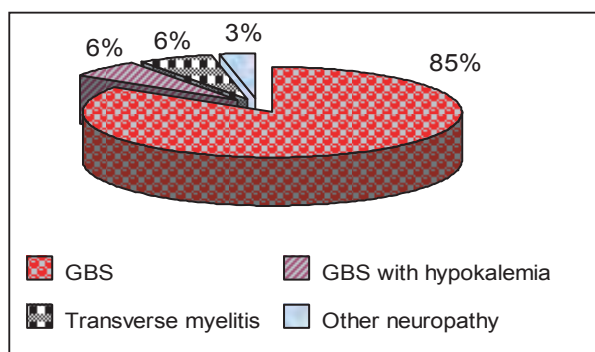


Fig 1: Different types of AFP

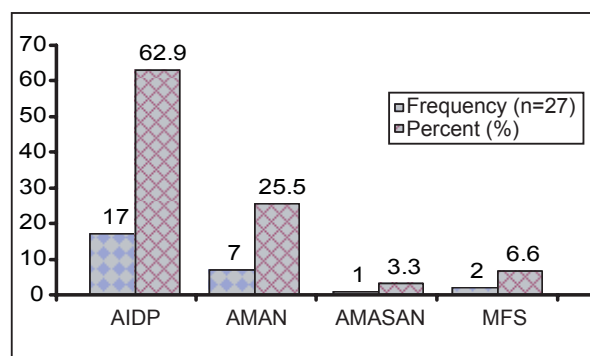


Fig 2: Types of GBS

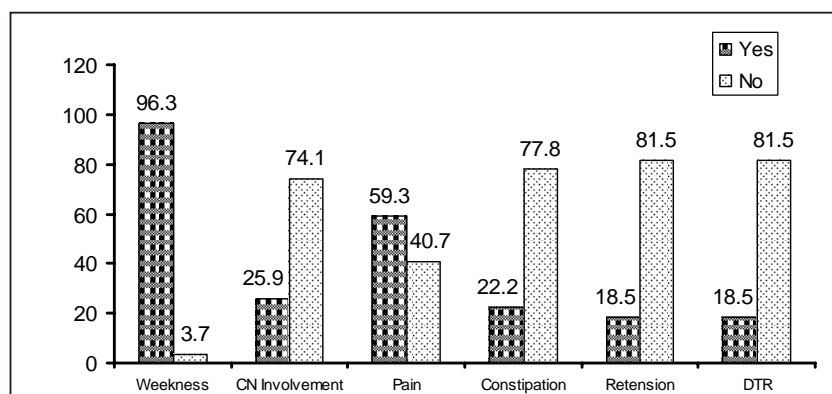


Fig 3: Clinical Presentation

Discussion

GBS is commonest cause of AFP, AIDP being commonest subtype in our setting. There was associated preceding illness in GBS. Majority patients presented with symmetrically ascending paralysis with gradual onset involving all limbs. Male were more affected in present study with male female ratio as 1.7:1.0. There was no significant age distribution but slightly higher patient from pre adolescent to young adults were found. There was no seasonal variation, no significant status with immunization, but 14.8% patients had relapse within 5 year. Patient had associated illness like URTI, pneumonia, sore throat, diarrhea in majority patients. Facial Nerve palsy was commonest cranial nerve involvement, in 60% of patients presented with sensory symptoms like pain, tingling and numbness. There was transient Bowel and bladder involvement in 1/5th of the cases. All patient became areflexia during the course of illness with 69.2% became bed ridden at nadir. There was albumino-cytological dissociation in 80% case. Majority patient improved with supportive treatment alone, 19.5% patient required ventilator support among them 40% mortality occurred. Majority of patient discharged within 1 week of admission with some improvement. 7.4% cases are expired during treatment, at 3 month follow-up almost half patient become healthy and remaining patient were improving on follow up. GBS is the commonest cause of the acute flaccid paralysis. It has been observed in Nepal from long time, which was recorded nationwide through AFP surveillance. Our present study was carried out to find out the clinico-epidemiological features of GBS in eastern part of Nepal (carried out at BPKIHS), the only tertiary care centre in Eastern Nepal.

Population based surveillance in Netherlands (1992-94)⁸ shows the incidence of GBS as 30.8% Idiopathic Neuropathy with AFP 12% and infection related AFP 40%. AIDP was commonest of all types of GBS. In western countries, AIDP was highest; where as in Northern China AMAN was the highest incidence. Rees JH, et al (1995), England study⁹ shows 85% AIDP and MFS 5%¹⁰. Ho TW, et al in Western China study¹¹ show AIDP – 19%, AMAN 65%, AMASAN 10-15%. Udaya Seneviratne, Srilanka Study (2000)¹² shows, 76% AIDP, 24% AMAN and 10% comprises other subtype of GBS which was comparable to our study. Rong KL, et al (1997)¹³ Taiwan study showed AIDP (49%), AMAN (4%), MFS (18%) and others 29%. Emilia Romagna, Italy study (1992-93)¹⁴ showed AIDP (36%), AMAN (14%) and MFS (13%). As shown in most of the population based studies our study had shown highest number of AIDP patients.

Rees JH, et al (1998) in south east England study¹⁵ had shown slightly predominant in adolescents and

young adults. Beghi E, et al (1985) Netherlands study¹⁶, Larsen JP, et al (1985) Norway study, Vankoninsveld R, Netherlands study (2000)¹⁷ show there was linear increase in incidence with age. In this study though there was no specific age variation, most patients were between 11-15 years (40%) of age. Netherlands⁸ study shows no consistent seasonal variation. But Emilia-Romagna study group Italy (1992-93)¹⁴, Beghi E, et al (1985) study, rural northern China study show relatively higher incidence in summer months, which was associated with increased infection with campylobacter infection. Dana I et al study shows 2/3rd children had associated illness like fever (59%) and diarrhea (26%). In Emilia Romagna study in Italy (1999)¹⁴ 65% cases had associated infection (59% fever, 18.2% diarrhea); Cheng Q, et al Sweden study (2000)¹⁸ shows 60% respiratory tract illness, 28% diarrhea, 45% RTI. Paul H, et al (2001)¹⁹ study show 71% associated illness, Udaya Seneviratne, Srilanka study (2000)¹² show 60% associated illness (20% Diarrhea, 40% Respiratory tract infection), Nicholas DL, et al²⁰ study shows 71% associated illness, Taiwan study (1997) show 67% associated illness (54% respiratory infection). Jacob BC, et al Netherlands study (1998)²¹ show 32% diarrhea and 19% respiratory tract illness. In this study, 73.5% patients presented with associated illness. 66.7% patient with preceding history of fever within 1 week, 22.2% patients with diarrhea, and 41.8% respiratory illness.

Udaya Senevirante (2000)¹² shows 53% CN weakness, Emilia Romagna Italy study (1992-93) 53% Spain study (1994) by Sedano MJ, et al shows 50% of Cranial Nerve weakness. In this study 25.5% patient shows Cranial Nerve weakness, facial nerve weakness being the commonest. In present studies lesser number of cranial nerve involvements was found as compared to others. Mortality rate ranges from 2-13% in different studies. Dana I, et al²² study shows 5%, Italian GBS study group shows (up to 12%), Cheng Q, et al (2000)¹⁸ shows 8%, Nicholas, et al (1998)²⁰ shows 4%, Italian study (1988-93) 9%, South England (1993-94) shows 9%, Voston MGH (1962-81) shows 2%, Brazil study (2003) shows 10% mortality among all GBS case. In our study mortality rate was 7% which is comparable to other studies. In our study majority of patients recovered over 1 year period. At 3 months follow-up shows 43.47% became healthy and 9% bedridden. Italian GBS study group shows 15% bed ridden, where as Chang Q, et al (2000) study shows 4% bed ridden at 3 month follow-up.

Conclusion

GBS is the commonest cause of AFP, AIDP being commonest subtype in our setting. We have to improve our existing treatment facilities and extend to different centers to detect and treat GBS. Most of the patients

improve with supportive treatment alone. Ventilator support indicates grave prognosis. Adequate supportive treatment and long term follow up is needed to insure recovery from illness. Over all cures is good except respiratory failure.

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Conflict of Interest

This article is based on the final thesis (part of compulsory thesis submission) report submitted to Department of Paediatrics and Adolescent Medicine, BP Koirala Institute of Health Sciences, Dharan.

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