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Experience With Guillain Barre' Syndrome (GBS) in Children

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SUMMARY

Guillain Barre' syndrome (GBS) is an acquired disease of peripheral nervous system. The precise etiology is not known and there is no distinctive test to confirm the diagnosis of GBS. All cases of GBS admitted in the department of paediatrics, Shaikh Zayed Hospital, Lahore, Pakistan during the last four years (1994-1997) were included in this retrospective study. Fourty cases of GBS were included in the study, 28 (70%) were male and 12 (30%) were females with a M:F ratio of 2.3:1. The age range of the patients was 1 year to 13 years with a mean age of 5.4 years. Twenty four (60%) of patients had both upper and lower limb involvement, while 16 (40%) had isolated lower limb involvement. Twelve (30%) had autonomic instability, 11 (27.5%) had respiratory muscle involvement and 1 patient developed transverse myelitis. Nineteen (47%) patients were not given any specific treatment such as steroids or immunoglobulins while in 16 (40%) high dose immunoglobulins were used. Corticosteroids were given in 5 (13%) patients. Four (10%) patients expired despite mechanical ventilation primarily due to autonomic disturbances. GBS is a common problem in children, the outcome is generally good even without any specific treatment, except when there is autonomic or respiratory muscle involvement.

INTRODUCTION

GBS is the most common cause of acute generalized paralysis at all ages[1]. The disease occurs in both sexes and at all ages and has been reported worldwide.The acquired nature of the disease, the response to immunotherapy and the pathology, all suggest that GBS is an immune mediated disease occurring in children after a viral infection.There is no distinct clinical presentation nor any laboratory test specific for this disorder. Furthermore, variants of the disease have emerged to make GBS an active syndrome rather than a specific disease[2-4].

The estimated incidence of GBS is approximately 0.4 to 1.7 cases per 100.000 population. The disease is rare in children below the age of one year[3]. Prognosis in children is worst if progression is prolonged and the onset of recovery is delayed[5]. Supremacy of immunoglobulins in the treatment of GBS has been documented by many authors. In some centres plasmapheresis has been reported to be a better modality of treatment in GBS, but it is not freely available[6]. We have done a retrospective study in 40 patients of GBS and have analyzed their clinical presentations, response to different treatments and their prognosis. In addition all patients were followed up for a period of one year to detect any residual damage, like motor deficit, after one year.

PATIENTS AND METHODS

Patients selection

All cases of GBS admitted in the department of paediatrics, Shaikh Zayed hospital, Lahore, Pakistan during the last four years (from 1994 to 1997) were included in this retrospective study. Patients of both sexes and all ages upto the age of 13 years were included.Those patients in whom the clinical diagnosis was in doubt and/or the laboratory findings were not conclusive of GBS were excluded from the study.

Those patients who died within six hours of

hospitalization or those who improved completely within 48 hours were not included in the diagnosis of GBS.

Investigations

Complete blood examination and cerebrospinal fluid (CSF) examination were performed in all patients. In CSF, protein estimation, total and differential cell counts and culture were done.

Patients with signs and symptoms suggestive of any imtracranial pathology were subjected to computerize axial tomographic (CAT) scanning. Nerve conduction studies were performed in patients with atypical presentation. Nerve conduction studies were also done in patients who had motor and / or sensory deficit, 6 months after discharge from the hospital.

Any patient suspected to be having transverse myelitis was confirmed by magnetic resonance imaging (MRI) scan.

Treatment modalities

Three treatment modalities were used in these patients:

1. **Supportive care:** No specific treatment and supportive care only was given to mild to moderately severely ill patients.

2. **High dose immunoglobulin therapy:** It was used in seriously ill patients or those having a rapid progression of the disease. Immunoglobulins were given in a dose of 400 mg/kg/day intravenously for 5 days.

3. **Corticosteriods:** In chronic patients or serious patients in whom immunoglobulis could not be arranged due to financial reasons, methylprednisolone was given in the dose of 5 mg/kg/day for two weeks.

All the patients were followed up in the out patient department and were discharged from follow up on complete recovery with no motor or sensory deficit on two consecutive visits at an interval of 4 to 8 weeks.

RESULTS

A total of 5417 patients were admitted in the Department of paediatrics ,Shaikh Zayed hospital during the four years of study (from 1993 to 1997). Out of these, 705 (13.2%) patients had disorders of the nervous system and amongst these 40 (5.67%) were diagnosed as cases of GBS (Table 1). Out of these 40 patients, 28 (70%) were males and 12 (30%) females with a male to female ratio of 2.3:1. The age range of the patients was 1 to 13 years, with the mean age of 5.4 years.

Table 1: Number of cases of GBS amongst patients admitted in the Deptt. of Paediatrics, SZH, Lahore (1994-1997).

Admissions	*Number*	*Percent*
Total admissions	5417	
Patients other than nervous system involvement	4712	86.98
Patients with nervous system involvement	705	13.01
Patients with GBS	40	5.67

Weakness of at least two limbs was observed in 100% of the patients. Sixty percent of these patients had involvement of both upper and lower limbs while 40% (16) of patients had involvement of lower limbs only, upper limbs being spared. In 7.5% (3) of the patients severity was more marked in upper limbs as compared to lower limbs. Involvement of respiratory muscles was observed in 11 (27.5%) of the patients.

In one patient despite the involvement of respiratory muscles, cranial nerves and both the upper and lower limb tendon reflexes were easily elicitable and the patient improved. Brain CT scan of this patient was normal.

Pain or paraesthesia was observed in 32.5% (13), whereas in 40% of the patients there was a loss of minimal response to painful stimuli. Autonomic involvement was observed in 12(30%) of the patients while bladder dysfunction was observed in 13% (5) and cardiovascular disturbances were observed in 10% (4) of the patients.

Three treatment modalitieswere tried in the patients of GBS (Table 3). These included high dose immunoglobulin therapy (400 mg/kg/day), supportive care only and steroid therapy (methylprednisolone in the dose of 5mg/kg/day).

Table 2: Clinical presentation of patients of GBS (n=40).

Clinical features	*Number*	*Percent*
Motor		
Muscular weakness	40	100
Upper and lower limb weakness	24	60
Isolated lower limb involvement	16	40
Upper limb involvement more than lower limb	3	7.5
Respiratory muscle paralysis	11	27.5
Intact tendon reflexes	1	2.5
Cranial nerve involvement	10	25
Sensory		
Pain or paraesthesia	13	32.5
Autonomic involvement	12	30
Bowel and bladder dysfunction	5	13
Cardiovascular disturbances	4	10

Table 3: Treatment modalities used in cases of GBS (n=40).

Type of treatment	*Number*	*Percent*
General supportive care	19	47
I/V immunoglobulins*	16	40
Corticosteroids**	5	13
Total	40	100

*High dose immunoglobulin therapy, 400mg/kg/day intravenously for 5 days.

**Methylprednisolone, 5 mg/kg/day for 2 weeks.

Immunoglobulins were used in 40% (16) of the patients while in 19 (47%) of the patients only supportive care was provided. Immunoglobulins were not used in all the patients due to financial constraints. In 5 (13%) of the patients, methylprednisolone along with general supportive care was used. These were either serious patients or chronic patients, but immunoglobulins could not be arranged for them. Due to non-availability of plasmaphresis, this form of treatment was not offered.

Thirty five patients were discharged in satisfactory condition but one patient developed transverse myelitis, upto the level of T12. The diagnosis was confirmed by MRI scans. Bowel and bladder incontinence was observed in this patient. There was little improvement in this patient after six months of discharge from the hospital (Table 4). Four patients expired during their stay in the hospital. All these patients had paralysis of the respiratory muscles and were on ventilator. The direct cause of death was cardiac dysrythmias.

On follow-up none of the patients reported with relapse although four patients were still having motor deficit after one year of discharge and all of these were ventilated for more than seven days during their stay in the hospital.

Table 4: Outcome of patients of GBS (n=40).

Outcome	*Number*	*Percent*
Discharged in satisfactory condition	35	87.5
Discharged with transverse myelitis	1	2.5
Motor deficit after one year of discharge	4	10
Expired	4	10

DISCUSSION

GBS is the most common cause of acute neuromuscular paralysis, yet its cause and pathogenesis are unknown[1]. It is clinically defined as a peripheral neuropathy causing limb weakness that progresses maximally up to 4 weeks before reaching a plateau. The annual incidence of GBS in Pakistan is not known. In our study 0.7% of the patients admitted during a four year period were diagnosed as cases of GBS.In the developed countries the estimated annual incidence of GBS is 2.4 to 1.7 cases per 100,000 paediatric population[2-4]. Shaikh Zayed hospital is a tertiary care unit, our rate of diagnosis of cases of GBS therefore may be higher as compared to other hospitals.In our study, out of 40 patients, 79% were males and 30% were

females. Hung et al 1994 have reported that males affected by this disease are about 61%[7].

The diagnosis of GBS is made by typical and atypical presentations. In our study all the patients had weakness and hyporeflexia except for one patient in whom the tendon reflexes were easily elicitable and the patient recovered completely.Weakness of all four limbs was observed in 60% of the patients, weakness was confined only to lower limbs in 40% of the patients, whereas arms were weaker than legs in 7% of these patients (Table 2). None of the patients had isolated upper limb involvement, so legs were involved in 100% of the patients. Involvement of upper limbs has been seen in 90% and arms weaker than legs in 5% of patients in a similar study[8]. Respiratory muscle paralysis requiring ventilation was recorded in 26%, sensory involvement was evident in 33% and autonomic involvement was recorded in 18% of patients in our study. Hung et al 1994, have reported sensory complaints in 26%, respiratory failure requiring ventilation in 14% and autonomic dysfunction in 46% of the patients[7]. The lower incidence of involvement of the autonomic nervous system in our study cannot be explained.

Three modalities of treatment were tried in the present study. It was a general observation in our study that immunoglobulins stop the progression of the disease, and the recovery is rapid, resulting in a shorter period of hospitalization as compared to patients in whom immunoglobulins are not used. Benefits of immunoglobulins have been demonstrated by many centres[8-12]. In 13% of our serious and chronic patients methylprednisolone was used. No clear benefit of this treatment was demonstrated over isolated supportive care in our study, the same results have been reported by Visser et al[10].

In the present study 39 (89%) of the patients were discharged in a satisfactory condition, 4 (10%) patients expired during stay in the hospital and 1 patient developed transverse myelitis.

Out of the patients who expired, two were receiving immunoglobulins, while two were on supportive care and methylprednisolone. All these four patients were neurologically very serious patients, and were on ventilator for respiratory failure. Another risk factor in these patients was involvement of the autonomic nervous system. All these patients died of cardiac dysrhythmias and hypotension. The same risk factors have been observed by Visser et al[10].

Although supremacy of plasmapheresis over immunoglobulin therapy has been described[13], due to non-availability of this treatment modality, it was not offered to our patients.

On follow-up four patients, in addition to the one having transverse myelitis had motor deficit even after one year of discharge from the hospital. However relapse did not occur in any of the patients.

It is concluded that rapid progression of the disease requiring ventilator support for a prolonged time and involvement of the autonomic nervous system are the risk factors for morbidity and mortality in GBS patients. Transverse myelitis has the worst prognosis in terms of recovery[13-14].

Regarding treatment, immuno-globulins have a better efficacy if provided earlier in the disease along with good general supportive care. Methylprednisolone seems to have no role in the treatment of acute GBS.

REFERENCES

1. Ropper AH. The Guillain - Barre syndrome. N Engl J Med 1992; 326: 1130-36.
2. Rantala H, Cherry ID, Shields WD, Uhari M. Epidemiology of GBS in children relationship of oral polio vaccine administration to occurrance. J Pediatrics 1994; 124: 220-30.
3. Evans OB. Guillain - Barre syndrome in childhood. Pediatr Review 1986; 8: 69-74.
4. Owen B. Evans and Vettaikoruniakan Kav Vddana Rayanan. Guillain - Barre syndrome. Pediatric Review 1997; 18: 10-16.
5. Hughes RA, Rees JH. Clinical and epidemiologic features of GBS. J Infect Dis 1997; 176: S92-S98.
6. Farkkila M, Kinnunen E, Weekstorm P. Survey of Guillain - Barre syndrome in southern Finland. Neuroepidemiology 1991; 10: 236-41.
7. Hung KL, Wang HS, Liou WY, Mak SC, Chi CS, Shen EY. Guillain - Barre syndrome in children: a cooperative study in Taiwan. Brain Dev 1994; 16: 204-8.
8. Shahar E, Shore, Roifman CM, Levi Y, Brand N, Ravid S, Murphy EG. Immunoglobulins are effective in severe pediatric Guillain - Barre syndrome. Pediatric Neurol 1997; 16: 32-36.
9. Reisin RC, Pociecha J, Rodriquez E, Massaro ME, Arroyo HA, Fejerman N. Severe Guillain Barre syndrome in childhood treated with human immune globulin. Pediatr Neurol 1996; 14: 308-12.
10. Sarada C, Inark K, Naim. Guillain - Barre syndrome: A prospective clinical study in 25 children and comparison

with adults. Ann Trop Paediatr 1994; 14: 281-86.
11. Visser LH, Van Der Meche FG, Meulstee J, Van Doorn PA. Comment in: J Neurol Neurosurg Psychiatry 1998; 64: 148-149.
12. Abd Allah SA, Jansen PW, Ashwal S, Perkir RM. Intravenous immunoglobulin as therapy for pediatric Guillain - Barre syndrome. J Child Neurol 1997; 12: 376-80.
13. Jansen PW, Perkin RM, Ashwal S. Guillain - Barre syndrome in childhood. natural course and efficacy of plasmapheresis. Pediatr Neurol 1993; 9: 16-20.
14. Singh NK, Jaiswal AK, Misra S, Srivastava PK. Prognostic factors in Guillain - Barre syndrome. G Assoc Physician India 1994; 2: 777-79.

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