ORIGINAL ARTICLE

Clinical Spectrum of Guillain-Barre Syndrome (GBS) in Children

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ABSTRACT

Objectives: To evaluate the clinical course of GBS in children.
Materials and methods: The study was conducted at Department of Pediatrics, the Children Hospital and Services Hospital, Lahore from 2001 – 2008 including 134 children up to the age of 15 years admitted with AFP. Routine investigations were done in all patients while CSF examination, nerve conduction studies (NCS) and electromyography (EMG) were done in most of the patients depending upon facilities available. Treatment modalities including supportive, intravenous immunoglobulin (IVIG) and plasmapheresis were used depending upon indications and facilities available.
Results: Out of 134 patients with AFP, 121 (90.3%) were diagnosed as GBS. Mean age of children with GBS was 6.2 years with a male to female ratio of 1.6:1. History of preceding illness was present in 67.8%, sensory symptoms in 71.9% and autonomic dysfunction in 61.1% patients. Classical GBS (symmetrical ascending paralysis) was present in 92.6% patients, 2.5% patients had descending paralysis, 4.1% had relapsing variety and one patient had chronic inflammatory demyelinating polyneuropathy. On electrophysiological studies, 33.3% patients had demyelinating variety, 44% had demyelinating with axonal involvement and 22.5% patients were labeled as unclassified. 53 (43.8%) patients required mechanical ventilation. 35.6% patients with axonal variety 23.5% patients with demyelinating variety required mechanical ventilation. 56 patients were treated with IVIG and out of these 36 patients improved, and 9 patients showed no improvement and plasmapheresis was done in these patients.
Conclusion: GBS is the commonest cause of AFP. GBS with axonal involvement is more severe as compared to demyelinating type and IVIG is an effective treatment modality for childhood GBS.
Key words: Acute Flaccid Paralysis, Guillain-Barre Syndrome, Children

INTRODUCTION

Guillain-Barre Syndrome (GBS) is an acute acquired demylenating polyneuropathy presumed to be immune mediated. It is the commonest cause of acute flaccid paralysis (AFP) after eradication of poliomyelitis[1,2,3]. An incidence of 0.5-5/100000 children/year has been reported worldwide[4,5,6].

GBS is characterized by muscle weakness and areflexia. The paralysis in GBS is flaccid and tends to be ascending and symmetrical with absent or diminished deep tendon reflexes (DTRs). Pain and muscle tenderness is prominent at the onset of paralysis. In addition to flaccid paralysis, the patient may also have sensory symptoms and involvement of cranial nerves, respiratory muscles and autonomic nervous system.

Diagnosis of GBS is usually clinical. In addition to routine investigations CSF examination and electrophysiological testing, nerve conduction studies and electromyography (NCS & EMG) are helpful for the diagnosis of GBS. Management of the child with GBS includes general measures like frequent monitoring, maintenance of airways and ventilatory support if indicated, feeding, bladder & bowel management and frequent change of posture. Specific management involves intravenous immunoglobulins and plasmapheresis. Steroids have no role in the management of GBS. Prognosis is good with complete recovery in more than 95% patients with GBS but it usually takes weeks to months[7].

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MATERIALS & METHODS

The study was conducted at Department of Pediatrics, The Children's Hospital and Services Hospital, Lahore from 2001 to 2008. A total of 134 patients up to the age of 15 years presenting with acute flaccid paralysis (AFP) were included in the study, while the patients with pseudoparalysis, CNS infections and encephalopathy, chronic flaccid paralysis and stroke were excluded.

All patients were admitted in pediatric ICU. Detailed history and clinical examination for distribution of weakness, cranial nerve involvement, sensory loss and autonomic failure was done in all patients. The patients were followed for progression of weakness, autonomic dysfunction and involvement

of respiratory muscles and bulbar paralysis. Routine investigations including CBC, ESR, serum electrolytes, and random blood sugar were done in all patients. CSF examination was done in the second week of illness. Nerve conduction studies and electromyography were done where possible. Treatment modalities including supportive, steroids, Intravenous Immunoglobulins (IVIG) and plasmapheresis were selected in patients with GBS depending upon indication and facilities available. Indications for IVIG were rapidly progressive disease, paralysis or impending paralysis of respiratory muscles, dysphagia and involvement of autonomic nervous system. Data was collected and results were analyzed using SPSS version.

RESULTS

A total of 134 children presenting with AFP were included in the study. Out of these, the largest group comprised of patients with GBS, 121 (90.3%). Other causes of AFP included transverse myelitis 6 (4.5%), poliomyelitis 2 (1.5%), traumatic neuritis 2 (1.5%) and one each patient of porphyria, periodic paralysis and renal tubular acidosis (RTA) (Table.1).

The clinical course of patients with GBS was further studied with reference to epidemiologic and clinical features, clinical variants and treatment modalities.

The age range of patients with GBS was 4 months to 14 years with mean age of 6.2 years. Out of these 6(4.9%) patients were <1 year, 47 (38.8%) were 1-5 years, 40(33%) were 6 - 10 years and 28 (23.1%) were 11 – 15 years (Fig.1). Seventy-four (61.2%) patients were boys with male to female ratio of 1.6:1**.** Peak incidence of GBS was observed during the months of July to September with a second peak during March and April (Fig.2).

History of preceding illness was noted in 82 (67.8%) patients. Respiratory tract infection was present in 29(24%), gastrointestinal tract infection in 27(22.3%), non-specific fever in 24 (19.8%), and rash in 2(1.7%) while 39(32.2%) patients had no preceding history of infection / illness. Duration of illness before admission was <2 days in 18 (14.9%) patients, 2 – 10 days in 88(72.7%) and >10 days in 15(12.4%). Weakness and areflexia / hyporeflexia involving legs were present in 100% of patients. Both arms and legs were involved in 106(87.6%), respiratory muscles in 55(45.5%) and cranial nerves in 42(34.7%) patients. Sensory symptoms were present in 87(71.9%) patients with pain in 80 (66.1%), parasthesias in 6(4.9%) and sensory loss in 1. Autonomic dysfunction was found in 74(61.1%) patients with hypertension and tachycardia in 32(26.4%), tachycardia alone in 28(23.1%), bradycardia in 8(6.6%), bowel and bladder disturbance in 30(24.8%) and diaphoresis in 35(28.9%) (Table-2).

Classical GBS (symmetrical ascending paralysis) was present in 112(92.6%) patients, 3(2.5%) patients had descending paralysis, 5(4.1%) children had relapsing variety and one patient had chronic inflammatory demyelinating polyneuropathy (Table-3). On the basis of electrophysiological studies (n=102), GBS was classified as demyelinating in 34 (33.3%) patients, demyelinating with axonal involvement in 45(44%) patient and unclassified in 23(22.5%) patients (Table-4). Cerebrospinal fluid examination was done in 101 patients and albuminocytological dissociation was found in 58(57.4%) patients. The criteria used for albuminocytological dissociation was CSF protein >80 mg/dl and cells <10/cmm.

Out of 121 patients, 53(43.8%) patients required mechanical ventilation. In this group (n=53), 8 patients had demyelinating variety, 16 had demyelinating plus axonal, 18 children belong to unclassified subgroup and in 11 patients electrophysiological studies could not be done due to nonavailability of bedside services for this study. Among ventilated patients (n=53) 35(66%) patients were successfully weaned off, 15(28.3%) children died and 3 patients left against medical advice (Table-5).

Treatment modalities including supportive, mechanical ventilation, steroids, Intravenous Immunoglobulins (IVIG) and plasmapheresis were used depending upon indications, severity of disease and availability. In the supportive group, out of 65 (53.7%) patients, 58(89.2%) patients improved, 5(7.6%) patients died and 2 patients showed no improvement. 56 (46.3%) patients were given IVIG. Out of these 36(64.3%) patients improved, 9(16.0%) patients showed no improvement, 7(12.5%) patients died and 3 patients left against medical advice. Out of 9 patients who were treated with plasmapheresis, 6 (66.7%) patients improved and 3 (33.3%) patients died (Table-6).

DSCUSSION

Acute flaccid paralysis in children is defined as acute onset of flaccid paralysis in one or more limbs or of bulbar paralysis in any child less than 15 years of age[1]. GBS is the commonest cause of AFP worldwide. Other causes include transverse myelitis, botulism, Tic bite paralysis and traumatic neuritis. Rarely it may be caused by polymyositis, diphtheria, porphyrias, drugs and toxins and Vit. B12 deficiency. In our study the commonest cause of AFP was also GBS followed by transverse myelitis, poliomyelitis,

and traumatic neuritis. Rare causes included porphyria, periodic paralysis and renal tubular acidosis. In a study from Australia common causes of AFP were GBS (47%) and transverse myelitis (19%) followed by acute disseminated encephalomyelitis, traumatic neuritis, tic bite paralysis and infantile botulism[1]. A study from Hong Kong showed GBS (42%) followed by T.M. (15%) as the two common causes of AFP in children[2]. Two different studies from our country also describe GBS as the leading cause of AFP[8,14]. In this respect out findings are consistent with the quoted studies. Poliomyelitis is still being reported in our country as is the case in our study, but it has been completely eradicated from all developed countries. We did not find any case of tic bite paralysis and infantile botulism in contrast to a study from Australia. Possibly the specific wood tic is not found in our settings.

Among children with GBS 53(45%) were under 5 years of age. It could be due to high incidence of infections in young children which is consistent with another study from Hong Kong and Central America[2,9]. Male to female ratio in our study was 1.6:1 which is again in confirmation with a study from Malaysia where this ratio was 1.3:1 [(10)]. With reference to seasonal distribution of GBS, two peaks were observed in our study. First peak was found in summer (July – September) and second in spring (March - April). These peaks could be explained on the basis of high incidence of gastrointestinal and respiratory tract infections in summer and winter respectively.

Involvement of respiratory muscles was present in 45.5% patients. It is comparable with a study from Pakistan in adults where this figure was 55.9%[11]. Cranial nerve involvement was found in 34.7% children which have been found to be 46% and 50% in other studies in pediatric patients[7,12]. Sensory symptoms and autonomic failure were noticed in 72% and 61% of children respectively. These results are comparable with a study in children where neuropathic pain and autonomic dysfunction was present in 79% and 51% of pediatric patients with GBS[7].

CSF albuminocytological dissociation was present in 57.4% of patients in our study while in another study it was found in 97.5% of patients[12]. We performed CSF examination in second week of illness in all patients and our criteria for dissociation was protein >80 mg/dl and cells <10/cmm. So this difference could be explained on the basis of timing, the CSF examination was done and dissociation criteria used by others.

On the basis of electrophysiological studies, we classified the children with GBS as demyelinating variety (33.3%), demyelinating with axonal involvement (44%) and unclassified (22.5%). In a study about childhood GBS, these ratios were acute inflammatory demyelinating (60.7%), acute motor axonal neuropathy (25%) and Miller Fisher Syndrome (12.5%)[13]. This discrepancy may be explained on the basis of variable immune response to preceding infections in different ethenic groups leading to different types of GBS.

Regarding electrophysiological type and need for ventilation, it was found that 23.5% (8 out of 34) in the demyelinating group, 35.6% (16 out of 45) in axonal variety and 78.3% (18 out of 23) in the unclassified group required ventilatory support. Mortality in children with axonal involvement was 31.3% as compared to 12.5% in demyelinating type. These observations support the fact that GBS with axonal involvement is more severe as compared to demyelinating type.

With reference to comparison of different treatment modalities, the patients who received intravenous immunoglobulins could not be compared with plasmapheresis group as plasmapheresis was done in patients who had already received IVIG and it could not be compared with the supportive group as the patients in whom IVIG was given have more serious disease as compared to supportive group.

In our patients with GBS mortality was 12.4%. All these patients belong to mechanically ventilated group with or without other treatment modalities. It shows that the patients who have severe disease at onset and required mechanical ventilation had poor prognosis. The high mortality in this group may also be related to complications of mechanical ventilation like infection and barotraumas.

Figure 1: Age distribution of patients with GBS (n = 121)

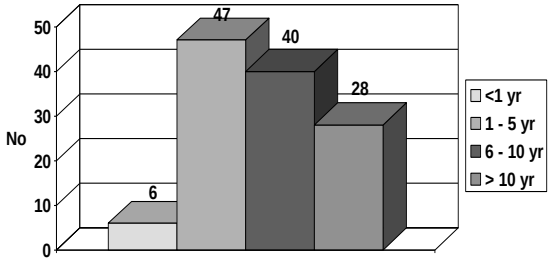

Figure 2: Seasonal distribution of GBS (n = 121)

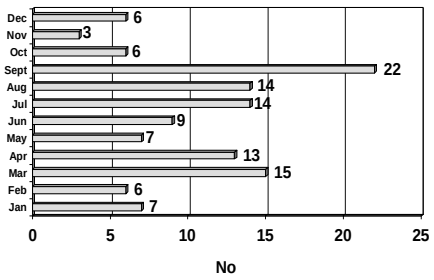

Table 1:Acute flaccid pralysis (n=1134)

Guillain barre syndreom	121
Trasverse myelitis	6
Traumatic neuritis	2
Prophyria	1
Periodic paralysis	1
Renal tubular acidosis	1

Table 2: Clinical presentation of GBS

Preceding H/O infection	
No H/O any illness	39(32.2%)
Non specific fever	24(19.8%)
GIT infection	27(22.3%)
RTI	29(24%)
Rash	2(1.7%)
Duration of illness prior to admission	
<02 days	18(14.9%)
2-10 days	88(72.8%)
>10 days	15(12.4%)
Weakness	121(100%)
Legs	121(100%)
Arms & legs	106(87.6%)
Respiratory muscles	55(45.5%)
Cranial nerves	42(34.7%)
Areflexia/Hyporeflexia	121(100%)
Sensory symptoms	87(71.9%)
Pain	
Parasthesis	6(4.9%)
Sensory loss	1(0.8%)
Autonomic symptoms	74(61.1%)
Diaphoresis	35(28.9%)
Hypertension & Tachycardia	32(26.4%)
Tachycardia	28(23.1%)
Bradycardia	8(6.6%)
Bowel & or bladder	30(24.8%)

Table 3: Clinical variants (GBS)

Variants	=n	%age
Classical GBS	112	92.6
Relapsing	5	4.1
Descending	3	2.5
Ch. Inflammatory demyelinating polyneuropathy	1	0.8

Table 4: Electrophysiological Studies (NCS & EMG)(n=102)

Types	**=n**
Demyelinating	34 (33.3%)
Demyelinating with axonal involvement	45 (44.0%)
Unclassified	23 (22.5%)

Table 6: Comparison of treatment modalities

Type	**No.**	**Improved**	**Not improved**	**Expired/ LAMA**
Supportive	65	58	2	5
MG	56	36	9	7+3
Plasmapharesis	9	6	-	3

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