ORIGINAL RESEARCH PAPER

Paediatrics

Volume - 7 | Issue - 4 | April-2017 | ISSN - 2249-555X | IF : 4.894 | IC Value : 79.96

Clinical Characteristics of Guillain-Barre Syndrome in children: A single center study

KEYWORDS

Abhay S Bagul

Associate Professor, Department of Pediatrics, Government Medical College, Akola, Maharashtra, India.

Vinit M Warthe

Associate Professor and Head, Department of Pediatrics, Government Medical College, Akola, Maharashtra, India.

Jaykumar Patel

Assistant professor, Department of Pediatrics, Government Medical College, Akola, Maharashtra, India.

ABSTRACT

Introduction: The Guillain-Barre syndrome (GBS) is an immune mediated acute polyradiculoneuritis and incidence of GBS ranges from 0.6 to 4.0/100,000 population. Now, GBS is most common cause of acute flaccid paralysis. Clinical characteristics of GBS in different regions and in different ages might be distinct. Hence, this study is planned to study clinical characteristics of GBS in children.

Methods: This was prospective observational study conducted from November 2014 to October 2016 and total 28 children enrolled in the study. A detailed history, clinical examination and cerebrospinal fluid examination of enrolled children was done and ten patients were treated with intravenous immunoglobulin.

Results: Out of 28, 15 (53.57%) were male and 13 (46.43%) were female. Age of subjects at time of presentation varies from 1 year to 12 years with mean of 5.89 and standard deviation ±3.02. Most common antecedent event was upper respiratory tract infection (25%). Occurrence of GBS was more in monsoon 16(57.14%). Motor weakness was most common presentation. Four patients had respiratory paralysis and out of four, two were died (7.14%).

Conclusion: Occurrence of GBS was more in monsoon (June to September) in our study. Motor weakness was most common presentation and nadir of weakness was within 7(4-14) days.

Introduction

The Guillain-Barre syndrome (GBS) is an immune mediated acute polyradiculoneuritis most frequently preceded by an unspecific infection [1]. The reported incidence rates of GBS range from 0.6 to 4.0/100,000 population. GBS described as a febrile generalized paralysis by Wardrop and Ollivier in 1834. Guillain, Barre and Strohl (1916) described a benign polyneuritis with albumin-cytological dissociation in the cerebrospinal fluid [2]. Asbury, Arnasonand and Adams (1969) described perivascular mononuclear inflammatory infiltration of the roots and nerves in GBS [3].

The disease is assumed to be autoimmune and operated by a preceding infection, mostly respiratory or gastrointestinal infections. Generally infections by microorganisms such as Campylobacter jejuni, CMV, Mycoplasma pneumonia, or influenza virus exist several weeks prior to approximately two thirds of GBS cases [4].

After eradication of poliomyelitis, GBS is the most common cause of acute flaccid paralysis. GBS usually presents as hypotonia, symmetrical motor weakness and hyporeflexia/areflexia and progressing to a nadir over a period of 4-6 weeks. The severity and duration of disease is highly diverse in patients and can range from mild weakness, from which patients recover spontaneously, to patients becoming quadriplegic and ventilator-dependent without signs of recovery for several months or longer. Clinical characteristics of GBS in different regions might be distinct due to geographical diversity and racial difference. Furthermore, genetic and environmental factors that affect an individual's susceptibility to develop the GBS are unknown [5]. Presentation and response to treatment of GBS is different in children and adults.

Hence, this study is planned to study clinical characteristics of GBS in children less than 12 years admitted in a tertiary care hospital.

Methods:

This was prospective observational study conducted from November 2014 to October 2016 in the Department of Pediatrics, Government Medical College and Hospital, Akola, Maharashtra, India. The clinical criteria proposed by Asbury and Cornblath used for diagnosis of GBS (Table 1).

Table 1: Diagnostic criteria for Guillain-Barre syndrome after Asbury and Cornblath [6]

I. Features required for diagnosis
(A) Progressive motor weakness of more than one limb
(B) Loss of tendon jerks
II. Features strongly supportive of the diagnosis
(A) Clinical features
1. Progression over four weeks
2. Relative symmetry of weakness
3. Mild sensory symptoms or signs
4. Cranial nerve involvement
5. Recovery, usually beginning two to four weeks after progression stops
6. Autonomic dysfunction
7. Absence of fever at the onset of neuritic symptoms
(B) Cerebrospinal fluid features
1. CSF protein raised after the first week of symptoms
2. Counts of 10 or fewer mononuclear leucocytes/mm 3
(C) Electrodiagnostic features
Reduction of conduction velocity, conduction block or abnormal temporal dispersion, increased distal latency or abnormal F wave in more than one nerve
III. Features casting doubt on the diagnosis
(A) Marked, persistent asymmetry of weakness
(B) Persistent bladder or bowel dysfunction
(C) Bladder or bowel dysfunction at onset
(D) More than 50 mononuclear leucocytes/ mm3 in CSF
(E) Presence of polymorphonuclear leucocytes in CSF
(F) Sharp sensory level
IV. Features that rule out the diagnosis
(A) Indication of any metabolic, infectious, or toxic disease associated with Polyneuropathy
(B) Occurrence of a purely sensory syndrome

Volume - 7 | Issue - 4 | April-2017 | ISSN - 2249-555X | IF : 4.894 | IC Value : 79.96

Total 28 children, 15 male and 13 female, less than 12 years were enrolled in the study after diagnosis of GBS. The study was approved by the institutional ethics committee and a written informed consent was obtained from parents of all study participants.

A detailed history, clinical examination and cerebrospinal fluid examination of enrolled children was done as per the pre-structured proforma at our hospital. All enrolled subjects were evaluated for presence of antecedent infection or event like vaccination. Detailed neurological examination for cranial nerve involvement, motor system, sensory system and autonomic dysfunction was done. Medical Research Council (MRC) sum score was used for evaluating the muscle strength from 0 to 5 in proximal and distal muscle in upper and lower limb bilaterally. Ten subjects were treated with intravenous immunoglobulin (IVIG). Nerve conduction velocity (NCV) was done in 8 children only due to scarcity of resources. Out of 28 children, two were died due to respiratory failure. Stool sample was collected and sent for analysis in all subjects according to AFP (acute flaccid paralysis) surveillance programme. All subjects were monitored for respiratory insufficiency and mechanical ventilation was provided in the presence of any of factors like clinical evidence of the use of accessory muscles, presence of severe bulbar weakness with risk of aspiration, arterial blood gas showing pO2<70 mm Hg or pCO2>45mm Hg.

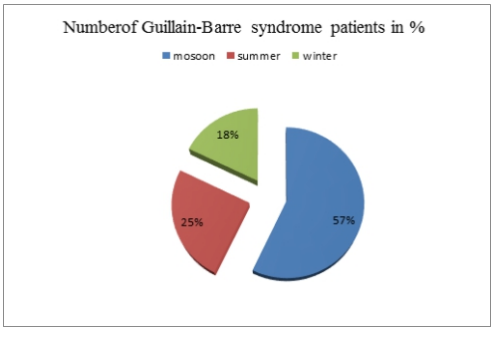
Numerical data presented as median, average and percentages.

Result:

Total 28 children were enrolled in the study during November 2014 to October 2016. Out of 28, 15 (53.57%) were male and 13 (46.43%) were female. Age of subjects at time of presentation varies from 1 year to 12 years with mean of 5.89 years and standard deviation ±3.02. Fever was present in one subject at the onset. Antecedent event before the weakness were found in 14 (50%) subjects and most common was upper respiratory tract infection (7, 25%) followed by viral infection and diarrhea.

Occurrence of GBS was more in monsoon (June to September) 16(57.14%), followed by summer (February to May) (7, 25.00%) and winter (October to January) (5, 17.85%) (Figure 1).

Figure 1: Seasonwise distribution of cases of Guillain-Barre syndrome



Neurological involvement at time of admission and at nadir is described in Table 2.

Table 2: Clinical characteristics of children with GBS (n=28)

Variables	Variables	Frequency	Percentage
Demography	Male/female ratio	1.15:1	_
	Age (Years)	5.89 ±3.02	_
Symptoms of antecedent infection	Upper respiratory tract infection	7	25.00
	Diarrhea	2	7.14
	Viral infection	5	17.85
Neurological symptoms at entry	Cranial nerve involvement	4	14.28
	Symmetrical weakness	26	92.85
	Asymmetrical weakness	1	3.57
	Sensory deficit	7	25.00
	Autonomic dysfunction	4	14.28
	Bladder involvement	2	7.14
	Pain	7	25.00
Neurological symptoms at nadir	Cranial nerve involvement	13	46.42
	Limb weakness	27	96.42
	Severe weakness (MRC 0-1/5)	12	42.85
	Respiratory paralysis	4	14.28
	Autonomic dysfunction	10	35.71
	Bladder Involvement	5	17.85
	Duration of Nadir	7(4-14)	_
Treatment	Intravenous immunoglobulin	10	35.71
	Plasmapheresis	0	0
	Symptomatic	18	64.28

Motor weakness was most common presentation. Cerebrospinal fluid examination was done in all patients in second week of illness or afterward. We found cell count less than 5/cumm in 20(71.42%), 5 to 10/cumm in 4(14.28%), 10-50/cumm in 2 (7.14%) and more than 50/cumm in 2(7.14%). Protein concentration was more than normal in all patients. Nerve conduction velocity was done in 8 patients and demyelinating pattern was found in 5 patients, axonal degeneration in 2 patients and mixed pattern in 1 patient. Four patients had respiratory paralysis and required ventilation assistance. Out of four, two were died (7.14%). We didn't find any case of acute relapse.

Discussion:

Total 28 subjects, from 1 year to 12 year, were enrolled in the present study and male to female ratio was 1.15:2. This slight difference in our study can be explained by extra attention and care given to male child during illness in our region. However, Dhadke SV et al found GBS more common in male with male to female ratio of 1.5:1 [7]. GBS can occur in all ages. Youngest patient in our study was of 1 year. In children, GBS is more common after 3 years [8]. A study By Sarkar et al reported youngest patient of 9 months age and GBS slightly more common in male [9]. Most common antecedent event was upper respiratory tract infection found in 50% patients. Guillain-Barré syndrome is described as a post-infectious disorder, as shown by the rapidly progressive, monophasic disease course (<1 month) shortly after infection, usually without relapse. Two-thirds of adult patients report preceding symptoms of a respiratory or gastrointestinal tract infection within 4 weeks of onset of weakness [10]. Widely studied antecedent infection is campylobacter jejuni infection. A study from France showed relation between GBS and recent influenza infection and more likely mechanism reported is immune mediated [11]. Similar to recent study in Maharashtra [12], we found more cases of GBS in monsoon season as compared to other seasons. This finding is contrary to study from Bangladesh, which found more cases in summer [13].This difference can be explained by influence of environmental factors and antecedent infections on etiology of GBS. High incidence of GBS also found after influenza vaccination [11]. It is believed that GBS is induced by both cell-mediated and antibody-mediated immune responses.

Most common presentation in our study was motor weakness. Similar study from India also found motor weakness as presentation in all enrolled patients (100%) [14]. Cranial nerve involvement was found in 14.28% patients on diagnosis and in 46.42% patients at peak of illness. Cranial nerve involvement is common in GBS in children and adult [14]. Duration of nadir of weakness from onset in our study was 7(4-14) days. Although, progression can occur up to 6 weeks from onset, nadir is usually seen within first 2 weeks in 80% of patients [15]. Two patients were died in our study due to respiratory paralysis. Other common cause of death in GBS is autonomic dysfunction. Overall prognosis of GBS is good in children [16]. Most children recover from weakness within six months; however continued

Volume - 7 | Issue - 4 | April-2017 | ISSN - 2249-555X | IF : 4.894 | IC Value : 79.96

improvement occurs beyond one year. As described by study from India, factors significantly associated with poor outcome at one year follow up were; artificial ventilation, inexitable nerves on nerve conduction and delayed independent walking [17]. On cerebrospinal fluid examination, we found albuminocytological dissociation.

We treated 10 (35.71%) patients with intravenous IG who presented with acute rapid progression of weakness and cranial nerve involvement. Plamapheresis not used in any patient. Intravenous IG is preferred because of easy administration and lower cost. Outcome after IVIG could not be measured in our study. As long term follow was not done we didn't comment on chronic inflammatory demyelinating neuropathy or sequela.

Conclusion:

Guillain-Barré Syndrome in children is more in male and age of presentation was 5.89 years and standard deviation ±3.02. Occurrence of GBS was more in monsoon (June to September) in our study. Motor weakness was most common presentation and nadir of weakness was within 7(4-14) days. Two patients (7.14%) were died.

References:

1. Korinthenberg R, Schulte Monting J. Natural history and treatment effects in Guillain-Barre syndrome: a multicentre study. Arch Dis Child 1996; 74: 281-7.
2. Guillain G, Barry J, Strohl A. Sur un syndrome de radiculonevrite avec hyperalbuminose du liquid cephalo-rachidien sans reaction cellulaire. Remarques sur les caracteres cliniques et graphiques des reflexes tendineux. Bull Mem Soc Med Hop Paris 1916; 40:1462-70.
3. Asbury AK, Arnason BG, Adams RD. The inflammatory lesion in idiopathic polyneuritis. Its role in pathogenesis. Medicine (Baltimore) 1969; 48:173-215.
4. Vriesendorp FJ, Mishu B, Blaser MJ, Koski CL. Serum antibodies to GM1, GD1b, peripheral nerve myelin, and Campylobacter jejuni in patients with Guillain-Barre syndrome and controls: correlation and prognosis. Ann Neurol 1993; 34:130-5.
5. Blum S, McCombe PA. Genetics of Guillain-Barre syndrome (GBS) and chronic infl ammatory demyelinating polyradiculoneuropathy (CIDP): current knowledge and future directions. J Peripher Nerv Syst 2014; 19: 88-103.
6. Asbury AK, Cornblath DR. Assessment of current diagnostic criteria for Guillain-Barre syndrome. Ann Neurol 1990; 27(suppl): S21-S24.
7. Dhadke SV, Dhadke VN, Bangar SS, Korade MB. Clinical Profile of Guillain Barre Syndrome. J Asso Physicians India 2013; 61: 168-72.
8. Van der Linden V, da Paz JA, Casella EB, Marques-Dias MJ. Guillain-Barre syndrome in children: clinical and epidemiological study of 61 patients. Arq Neupsiquiatr 2010; 68:12-17.
9. Sarkar UK, Menon L, Sarbapalli D, Pal R, et al. Spectrum of Guillain-Barré syndrome in tertiary care hospital at Kolkata. J Nat Sci Biol Med 2011; 2: 211-5.
10. Jacobs BC, Rothbarth PH, van der Meche FG, et al. The spectrum of antecedent infections in Guillain-Barre syndrome: a case-control study. Neurology 1998; 51: 1110-15.
11. Tardy VS,Orlikowski D, Porcher R, Sharshar T, et al. Guillain-Barré Syndrome and Influenza Virus Infection. Clin Infect Dis 2009; 45: 45-56.
12. Meshram RM, Bokade CM, Merchant S, Abhisheik S, et al. Clinical profile of childhood Guillain-Barre Syndrome. Int J of Pediatr Res 2016; 3: 427-32.
13. Islam Z, Jacobs B, Islam MB, Mohammad QD, et al. High Incidence of Guillain-Barré Syndrome in Children, Bangladesh. Emerg Infec Dis 2011; 17: 1317-8.
14. Dhadke SV, Dhadke VN, Bangar SS, Korade MB. Clinical Profile of Guillain Barre Syndrome. J Asso Physicians India 2013; 61: 168-72.
15. Fokke C, Berg B, Drenthen J, Walgaard C,et al. Diagnosis of Guillain-Barré Syndrome and validity of Brighton criteria. Brain 2014; 137:33-43.
16. Salehiomran MR, Nikkhah A, Mahadavi M. Prognosis of Guillain-Barré Syndrome in Children. Iran J Child Neurol 2016; 10:38-41.
17. Karla V, Sankhyan N, Sharma S, Gulati S. Outcome in Childhood Guillain-Barré Syndrome. Indian J Pediatr 2009; 76: 795-99.