Guillian Barre syndrome epidemiology presentation and outcome

By
Dr.Raed jabbar Hussain[*1]; Dr. Rahan Assim Mohammed Al-Qazzaz[*2]; Kahtan Adnan Abdullah[*3]
[*1] FIBMS Pediatrics, Al-Elwiya pediatric teaching hospital, Baghdad- Alrusafa/Health Directorate, Ministry of Health and Environment, Baghdad, Iraq; [*2] CABMS pediatrics, alsheikhan general hospital; [*3]Senior pediatrician F.I.B.M.S,JAR pediatric teaching hospital Suliamania

Abstract:

Background Guillain Barre syndrome (GBS) is most common cause of acute flaccid paralysis, affect peripheral nerves with distinctive features clinical, pathological and prognosis. **Patient and methods** this study is prospective of 60 patients admitted at al-kadymia teaching hospital from first January 2004 to end April 2009. Age includes 1 to ≤11 years. Diagnosis was by clinical examination and confirmed by CSF tests and nerve conduction velocity study. **Results** It was found that 32 patients were male and 28 were female. Cranial nerves involved in 30% of patients. Sensory symptoms found in 16%. CSF changes was seen in 85.5% patients. Antecedent events were found in 27 patients out of 60, 14 had history of upper respiratory tract infection 45%, 7 had gastroenteritis 11%, 6 patients had history of fever 3 weeks earlier 10% and 15 patients had complicated by respiratory failure managed by mechanical ventilators, During this study 4 patients had been died. **Conclusions:** Current study conclude about 45% of patients had history antecedent events in as upper respiratory tract infection , gastroenteritis and fever, so cerebrospinal fluid CSF cell in the majority of cases within normal range and mostly lymphocytes, Cranial nerves were affected in most patients without serious sequels so Steroid was not given to most of patients in our study without any significant effect on the course of disease , Hospitalization was range from 2 week to 4 week, Recovery was range from 4 to 12 week and Death rate was 6% .

Key words: Guillian barre syndrome (GBS); polyneuropathy; cerebrospinal fluid (CSF); steroids; intensive care unit (ICU).

Introduction:

Guillian Barre syndrome GBS is an acute idiopathic monophasic acquired inflammatory demyelinating polyradiculoneuropathy characterized by symmetrical progressive ascending weakness, areflexia, variable sensory complaints, and elevated cerebrospinal fluid CSF protein without pleocytosis. The disorder is thought to result from a Postinfectious immune mediated process that predominantly affect motor nerves [(1).] guillian Barre syndrome is the most common cause of acute flaccid paralysis in healthy infants and children [(2)]. It has an annual incidence of 0.6 to 2.4 cases per 100.000 population and occur at all ages and in both sexes [(3)].

The incidence is lower in children, 0.38 and 0.91 cases per 100.000 in two reports [(4,5)] . GBS occurs rarely in children younger than two years of age, but can occur in infants [(6,7)]. Males are affected approximately 1.5 times more often than females in all age groups [(1),] the pathologic changes depend upon the form of GBS. In acute inflammatory demyelinating polyradiculoneuropathy and Miller Fisher variant, a focal inflammatory response develops against myelin producing Schwann cells or peripheral myelin. Infiltration of the epineural and endoneural small vessels (mostly veins) by lymphocytes and monocytes causes segmental myellin degeneration throughout the nerve. The inflammation is more intense at the junction of the dorsal and ventral roots [(8)]. The demyelination blocks electrical conduction along the nerve. Axonal degeneration occurs as a secondary response; the extent relates to the intensity of the inflammatory response all myelinated nerves (motor, sensory, cranial and sympathetic) can be affected. The breakdown of the blood-nerve barrier at the dural attachment allows transudation of plasma proteins into the CSF [(1),]

The severity of GBS in children dose not correlate with long-term outcome. As many as 85 percent of children can be expected to have an excellent recovery. Approximately one Half of patients are ambulatory by six months, and seventy percent walk within a year after onset [9,10]. Abetter prognosis is associated with a gradual evolution of weakness [11]. Mortality is approximately 3 to 4 %, and usually is secondary to respiratory failure or cardiac complications [9]

Patients and Methods: Between (beginning of January 2004 to end of April 2009) sixty children with GBS were admitted to Al-kadymia teaching hospital in Baghdad or referred from other hospitals for further care or for RCU admission, the study is prospective. The variables for analysis in this study include (age, sex, duration of hospitalization, presentation, antecedent infection, motor weakness, sensory, autonomic and cranial nerves involvement, respiratory muscle involvement, admission to RCU, CSF analysis, and outcome). Investigation was done including lumber puncture (LP) in first 2 weeks, Electromyography (EMG) was done in 35 patients (58.3%). The criteria which had been applied in this study are the same diagnostic criteria of GBS after Asbury and cornblath includes the following (52). the **Features required for diagnosis**: Progressive motor weakness of more than one limb, Loss of tendon jerks .so f**eatures strongly support the diagnosis: Clinical features:** progressive over 4 weeks. Relative symptoms of weakness.; Mild sensory signs or symptoms. Cranial nerves involvement; Recovery usually beginning 2-4 weeks after progression are stops ; Autonomic dysfunction; Absence of fever at the onset of neurological symptoms; *CSF features:* CSF protein raised after the first week of symptoms .and counts of 10 or fewer mononuclear leukocytes ; ***Electrodiagnostic features:*** Reduction of conduction velocity, conduction

block or abnormal F wave in more than one nerve. ; **Features casting doubt on the diagnosis:** Marked persistent asymmetrical weakness; Persistent bladder or bowel dysfunction. ; Bladder or bowel dysfunction at onset ; Prescence of polymorphonuclear leukocytes in the CSF; Sharp sensory level; **Features that rule out the diagnosis:** Indication of any metabolic, infections or disease associated with polyneuropathy and Occurrence of a purely sensory symptoms.

Results:

About sixty children, 32 male and 28 females, age range between 1 - ≤11 years, half of them are 1 - 5 year of age, (table 1).

Table (1): Age and gender distribution.

Age	Gender				Total	
	Male		Female			
	No.	%	No.	%	Total	%
1 - 5 year	17	28.3	13	12.7	30	50
6 - 10	8	13.3	6	10	14	23.3
≤11	7	11.6	9	15	16	26.7
Total	32	53.3	28	46.7	60	100

about 27 patients (45%) had history of antecedent events (table 2). Upper respiratory tract infection (URTI) in (33.3%), gastroenteritis (11.7%) and 6 patients had history of fever, 33 (55%) of patients had no such events (table 2). 53 patients (88%) had a manifestation of typical GBS, 6 (10%) had Miller Fisher syndrome, 1 (1.6%) had atypical manifestations (table 2).

Table (2): Antecedent infection of patients with GBS and Clinical manifestations.

Types of previous infection	Number of patients	%

	(No.)	
Upper respiratory tract infection	14	33.3
Gastroenteritis	7	11.7
Fever	6	10
No history of infection	33	55
Total	60	100
Clinical form		
Typical GBS	53	88.3
Miller Fisher	6	10
Atypical	1	1.7
Total	60	100

Cranial nerves involvement was observed in 20 patients (30%). Motor nerves involvement in 60 (100%), Sensory manifestation was observed in 10 patients (16%) table 3.

Table (3): nerves involvement in 60 patients with GBS and sensory manifestation

Nerves	**Number of patients**	**%**
Cranial	20	30
Sensory	10	16
Motor	60	100
Sensory manifestation		
Paresthesia	7	11.7

Pain	12	20
No symptoms	41	68.3
Total	60	100

Autonomic nerves involvement was observed in 8 patients (table 4) as 3.3% (Urine retention; Stool and urine retention and Hypertension), but 86.7% No involvement (table-4).

Table (4): Autonomic nerves involvement.

Signs and symptoms	**Number of patients**	**%**
Urine retention	2	3.3
Urine inconvenience	1	1.7
Stool and urine retention	2	3.3
Cardiac dysrhythmias	1	1.7
Hypertension	2	3.3
No involvement	52	86.7
Total	60	100

CSF changes as increase protein was observed in (74%) patients, whilst only 26% as normal protein (table 5).

Table (5): CSF analysis and changes .

CSF examined	NO.	%
High protein	20	74
Normal protein	7	26
Total	27	100
CSF not examined	33	55

Four Patients (6%) died during acute illness because of respiratory muscles paralysis. 6 patients (10%) stayed in hospital for 2 weeks and 20

patients (33.3%) between 2 - 4 weeks, 53.3% patients for 5 - 8 weeks and only 2 patients (3.4%) more than 8 weeks, (table 6).

Table (6): duration of hospitalization and the outcome.

Time	NO.	%
1 - 2 weeks	6	10
2 – 4	20	33.3
5 - 8	32	53.3
More than 8 weeks	2	3.4
Total	**60**	**100**
Full recovery	50	83.3
Mild weakness	4	6.7
Death	4	6
Total	**60**	**100**

Full recovery occurred in 45 patients (75%) during 3-month period and 10 patients (16%) were discharged with mild weakness after 3 months, 4 patients (6%) are died (figure 1).

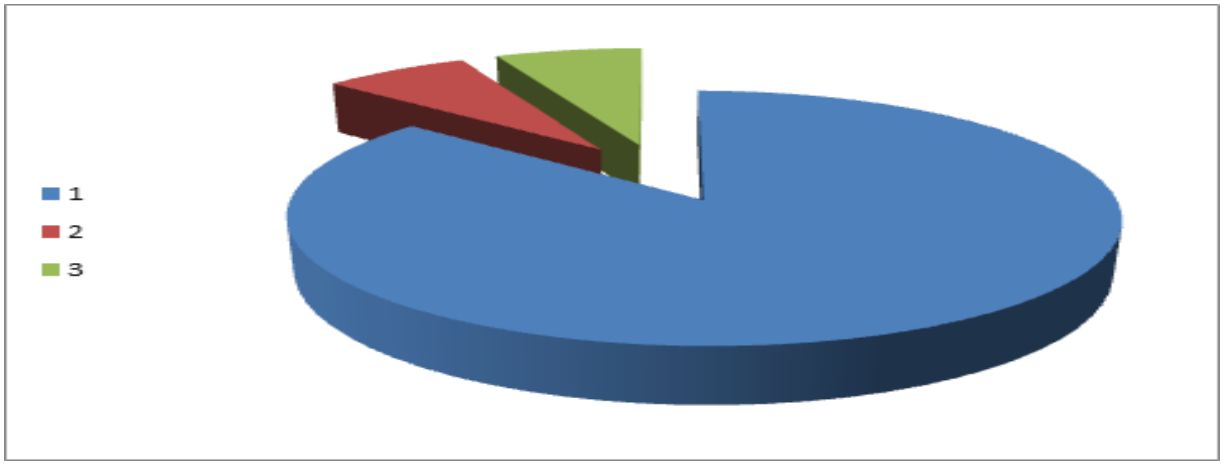

Figure (1): Outcome of patients with GBS (1. full recovery; 2. Mild weakness ;3. Death).

Discussion:

The age and sex of the patients were analyzed, it showed that male to female ratio 1- 1.1 this is in agreement with beghi *etal.*, which showed male predominance [3], so the age distribution in the majority of patients was 1 - 4 years and this in agreement with Hahn [12], Neurological manifestation were preceded by Antecedent events in 27 patients (45%), of these URTI (32.3%), gastroenteritis (11%) , fever in (10%) this is in regards to many studies [13 , 14 , 15] ,Ten patients 16 % had sensory symptoms as part of presenting clinical picture of GBS, while Osler [16] minimized the importance of sensory symptoms as part of presenting clinical picture of GBS.

Autonomic nerves involvements were recognized as part of GBS by birchfield [17], in our patients (15.4%) had evidence of autonomic dysfunction it was mainly in the form of urine incontinence and retention and cardiac dysrhythmias, however there is no prognostic significance of autonomic dysfunction and unrelated to the degree of paralysis, this is in agreement with [18] .

CSF protein was raised in 20 patients (85.5%), this is in agreement with rantala *etal.,* [4]. Highest value of CSF protein was (400mg/dl) and lowest was (20mg/dl). Normal CSF protein was found among 7 patients, may be related to the time at which LP were done, since protein content of CSF raises to its peak between 2^{nd}- 4^{th} week. Normal CSF protein especially early in the illness or finding of normal numerous lymphocytes less than 50/mm does not exclude diagnosis of GBS. [15]

In current study CSF contained up to 35 cell /mm, majority were lymphocytes, this in agreement with most authors who found that CSF may contain some cells and diagnostic criteria for GBS accepted cell count of less than 50 cell/mm [4,14,15]. In this study 4 patients died during acute illness giving death rate (6%), respiratory failure was found in 25% and these results are higher than that which was found in a study of Ropper [18].

Recovery over a period of weeks is a Hallmark of GBS and around 50 patients (83.3) have no residual deficit, 4 patients (6%) have persistent minor

problem such as foot drop which does not impair the conduct of everyday Life , this in agreement with Hahn[12]. Steroid (methyl prednisolone) was given to 5 Patients, it doesn't affect time of recovery or time of staying in RCU in comparison to those who had not been given and this was in agreement with some studies like that which had been done by Jones [2], so Plasmapheresis was done to only 1 patient with rapidly progressive course with dramatic response and not done to other patients because of unavailability of material and devices which is needed for plasmapheresis of patients' age. This is in agreement with Hahn [12].

References

1. Robert , P , Cruse , DO . Overview of Guillian Barre syndrome in children , http: up to date . Org. /.15.1_2007.
2. Jones ,HR Jr . Guillian Barre syndrome : perspective with infants and children . Semin pediatric neurology 2000 ; 7:91.
3. Beghi , E , Kurland LT , Mulder , DW wiederholt , WC . Guillian Barre syndrome . Clinicoepidemiologic features and effect of influenza vaccine . Arch neurol 1985 ; 42 : 1053 .
4. Rantala H , Uhari , M , Niemela , M . Occurrence , clinical manifestations , and prognosis of Guillian Barre syndrome . Arch Dis children 1991; 66 : 706 .
5. Olive , JM , Castillo , C , Castro , RG , de Quadros , CA Epidemiologic study of Guillian Barre syndrome in children less than 15 years of age in Latin America . J Infectious dis 1997 ; 175 suppl 1 : S160 .
6. Carroll , JE Jedziniak , M , Guggenheim , MA . Guillian Barre syndrome : another cause of the floppy infant . Am J Dis child 1977 ; 131 : 699 .
7. Buchwald , B de Baets , M , luijckx , GJ , Toyka , KV . Neonatal Guillian Barre syndrome : blocking antibodies transmitted from mother to child . Neurology 1999 ; 53 : 1246 .
8. Griffin , JW , Li , CY , Macko , C , et al. Early nodal changes in the acute motor axonal neuropathy pattern of Guillian Barre Syndrome . J Neurocytology 1996 ; 25 : 33 .

9. Evans , OB , Vedanarayanan , V . Guillian Barre syndrome . Pediatric Rev 1997 ; 18 : 10 .
10. Lu , JL , Sheikh , KA , Wu , HS , et al. Physiologic-pathologic correlation in Guillian Barre Syndrome in children . Neurology 2000 ; 54 : 33 .
11. Fisher , M . An unusual variant of acute idiopathic polyneuritis (syndrome of opthalmoplegia , ataxia and areflexia) . N Engl J Med 1956 ; 255 : 57 .
12. Hahn , AF . The Guillian Barre syndrome . Lancet 1998 ; 352 : 635
13. Patja , A , Paunio , M , Kinnunen , E , et al. Risk of Guillian Barre syndrome after measles-mumps-rubella vaccination . J Pediatric 2001 ; 138:250 .
14. Steininger , C , popow - Kraupp , T , seiser , A , et al. Presence of cytomegalovirus in cerebrospinal fluid of patients with Guillian Barre syndrome . J Infect disease 2004 :189:984 .
15. Mori , M , Kuwabara , S , Miyake , M , et al. Haemophilus influenzae infection and Guillian Barre syndrome . Brain 2000 ; 123 (pt 10) :2171.
16. Osler , LD and Heller : GBS N. Engl Med . 1960 , 262 : 964 - 969 .
17. 17.Birchfield , RI AND showe . Postural Hypotension in Guillian Barre Syndrome . Arch Neurol . 1964 ; 10 : 146-157 .
18. Ropper , AH . The Guillian Barre syndrome . N Engl J Med 1992 ; 326 : 1130 .