Clinical audit on the management of Guillain-Barre syndrome cases admitted to Assiut University Children Hospital

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Introduction
Guillain-Barré syndrome (GBS) is the most common cause of acute flaccid paralysis in children. It is characterized by progressive, symmetrical weakness, and areflexia in a previously healthy child. GBS should be diagnosed quickly in patients presenting with rapidly progressive paralysis. Meticulous monitoring and supportive care are needed in all GBS patients. Efforts focus on the follow-up of clinical course and outcome to improve the care and treatment of individual patients.

Materials and methods
The study included pediatric patients with GBS referred to Assiut University Children Hospital in 9 months from November 2016 to July 2017. Information was taken from resident doctors and mothers.

Results
Intravenous immunoglobulin (IVIG) was administered to 17 (80.95%) cases. Four (19.05%) cases who presented in the improving phase of motor disability were managed supportively and recovered completely. Plasmapheresis was done to three (14.3%) cases, who did not respond well to IVIG. Complete recovery was observed in 17 (80.95%) cases and four (19.05%) cases experienced only incomplete recovery.

Discussion
Nerve conduction velocity is very useful to diagnose GBS. IVIG and supportive care are considered key elements in the management of childhood GBS. IVIG administration early in the course of the disease results in better recovery. Plasmapheresis may be an effective option in cases with poor response to IVIG therapy.

Keywords:
Guillain-Barre syndrome, intravenous immunoglobulin and plasmapheresis, progressive paralysis

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Introduction

In 1916, Georges Guillain and Jean–Alexandre Barré published the very first comprehensive detailed classic paper on Guillain-Barré syndrome (GBS) [1]. The initial diagnostic criteria were published in 1981 and refined in 1990 [2,3]. GBS is an immune-mediated disorder of the peripheral nervous system, which is triggered by either infectious or noninfectious factors [4]. It is a rapid-onset muscle weakness caused by an autoimmune response damaging the peripheral nervous system. GBS is mostly preceded by infection or other immune stimulation inducing autoimmune response. Molecular mimicry between microbial and nerve antigens is obviously a major triggering factor behind the development of the disorder [5].

Genetic and environmental factors that affect an individual's susceptibility to develop the disease are unknown [6]. Abnormal autoimmune response does not develop in most individuals (>99%) exposed to an immune stimulus associated with GBS such as campylobacter jejuni [7]. Acute progressive limb weakness, often with sensory and cranial nerves affection, usually develops 1–2 weeks after immune stimulation and progresses to peak clinical disability in 2–4 weeks [8]. Nerve conduction velocity (NCV) is very useful to diagnose GBS. Many studies consider the treatment of choice to be intravenous immunoglobulin (IVIG) or plasmapheresis with supportive care. IVIG should be given for 5 days at a dose of 400 mg/kg/day [9].

Aim

The aim was to implement the use of the guidelines for management of GBS according to the American Academy of Neurology 2012 in Assiut University Children Hospital for 3 months and then audit the degree of adherence of the physicians to the recommended guidelines for 9 months.

Materials and methods

Methods

The study included all pediatric cases with GBS referred to Assiut University Children Hospital in 9 months. Information was taken from resident doctors and mothers.

Site of the study

Assiut University Children Hospital.

Duration of the study

A period of 9 months from November 2016 to July 2017.

Inclusion criteria

All cases presented with classic presentation of symmetrical ascending acute lower limb weakness, hypotonia, and hyporeflexia under the age of 18 years supported by NCV showing delayed F-wave and cerebrospinal fluid (CSF) examination in selected cases showing the characteristic cytoalbuminologic dissociation.

Exclusion criteria

(1) Cases of lower limb weakness due to causes other than GBS (Table 1).
(2) Cases of respiratory failure needing ICU other than GBS.

The diagnosis of GBS is mainly based on the clinical evaluation and exclusion of possible alternative diagnoses. We conducted a complete neurological examination on all cases. NCV was done in all cases.

CSF examination was done in selected hemodynamically stable cases by lumbar puncture. We observed all cases for respiratory insufficiency and we provided mechanical ventilation in the presence of any of these factors:

(1) Clinical use of accessory muscles.
(2) Respiratory muscle fatigue.
(3) Severe bulbar weakness with risk of aspiration.

Clinical course, results of electrodiagnostic studies, treatment given, and outcome were assessed.

Ethical consideration

Patients signed an informed consent.

IRB: 17100993.

Assiut Faculty of Medicine approved the study.

Statistical analysis

Data were analyzed and processed using the SPSS software (Chicago, Ill., USA), version 16 in the form of frequency mean ± SD, range, and percentages.

Results

A total of 21 children with GBS were admitted to our pediatric hospital during the period of the study, of which 11 (52.4%) were men and 10 (47.6%) were women (Fig. 1). Age of the patients ranged from 1.5 to 13 years with a mean of 6.6 ± 3.2 and majority of them were 7–10 years age group (42.8%) (Table 2). History of preceding infection was found in 13 (61.9%) cases. The main symptoms included weakness of upper and lower limbs, symptoms of cranial nerve affection, and symptoms of autonomic dysfunction, gait disturbance, dyspnea, fever, and muscular and radicular pain as shown in Table 3.

The main signs included hyporeflexia, hypotonia, signs of cranial nerve affection, signs of autonomic dysfunction, symmetry of weakness, ataxia, and sensation as shown in Table 4.

CSF analysis was performed in six (28.6%) cases and cytoalbuminologic dissociation was seen in only three (14.3%) cases. Nerve conduction studies were done and showed delayed F-wave in 19 (90.48%) cases and two (9.52%) cases were normal (Table 5).

IVIG was administered to 17 (80.95%) cases. Plasmapheresis was done to three (14.3%) cases, who did not respond well to IVIG. Four (19.05%) cases who presented in the improving phase of motor disability were given supportive care without IVIG or plasmapheresis and recovered completely (Table 6).

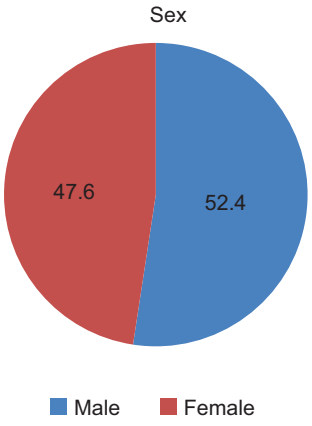
Respiratory insufficiency was found in six (28.6%) cases, requiring assisted ventilation (Table 7).

Complete recovery was observed in 17 (80.95%) cases and four (19.05%) cases experienced only incomplete recovery (Fig. 2). In our hospital, the American Academy of Neurology guidelines for management of GBS are being followed to a great extent and with meticulous care and attention resulting in a favorable outcome.

Discussion

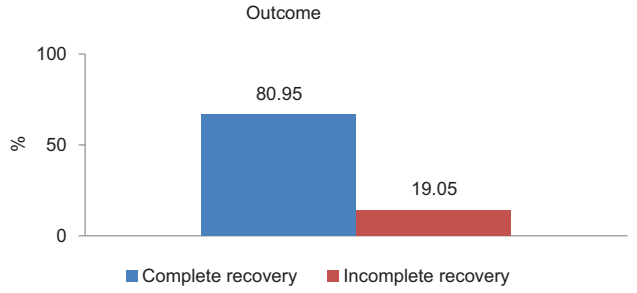
GBS is the most common cause of acute flaccid paralysis in children and infants after the eradication

Figure 1



Sex distribution among studied cases.

Figure 2



Outcome of studied cases.

Table 1 Differential diagnosis of Guillain-Barré syndrome

Disease	How to differentiate from GBS
Poliomyelitis	Fever, asymmetrical descending paralysis
Transverse myelitis	Sensory level, loss of bowel and bladder control, no cranial nerve affection
Myasthenia gravis	Normal tone and reflexes, no sensory affection, no cranial nerve affection
Botulism	Descending paralysis, oculobulbar affection
CNS infections	Convulsions, disturbed level of conscious

CNS, central nervous system; GBS, Guillain-Barre syndrome.

Table 2 Age groups of studied cases

	Range (mean±SD) and *n* (%)
Age (years)	1.5-13 (6.62±3.2)
Age group (years)	
<3	2 (9.52)
3-6	8 (38.09)
7-10	9 (42.85)
>10	2 (9.52)

Table 3 Symptoms of studied cases

Symptoms	Yes [*n* (%)]	No [*n* (%)]
History of preceding infection	13 (61.9)	8 (38.1)
Respiratory tract infection	5 (23.809)	
Gastrointestinal tract infection	4 (19.047)	
Nonspecific febrile illness (fever)	4 (19.047)	
Weakness of lower limbs	21 (100.0)	0 (0.0)
Weakness of upper limbs	17 (81.0)	4 (19.0)
Symptoms of cranial nerve affection		
Oculomotor	1 (4.8)	20 (95.2)
Facial	1 (4.8)	20 (95.2)
Bulbar	7 (33.3)	14 (66.7)
Symptoms of autonomic dysfunction		
Urinary incontinence	6 (28.6)	15 (71.4)
Stool incontinence	3 (14.3)	18 (85.7)
Disturbance in gait	18 (85.7)	3 (14.3)
Muscle pain	7 (33.3)	14 (66.7)
Radicular pain	4 (19.0)	17 (81.0)
Dyspnea	9 (42.9)	12 (57.1)
Fever	6 (28.6)	15 (71.4)

Table 4 Signs of studied cases

Signs	Yes [*n* (%)]	No [*n* (%)]
Hyporeflexia or areflexia	21 (100.0)	0
Hypotonia	21 (100.0)	0
Signs of cranial nerve affection		
Ophthalmoplegia	0	21 (100.0)
Facial palsy	1 (4.8)	20 (95.2)
Absent gag reflex	7 (33.3)	14 (66.7)
Ataxia	12 (57.1)	9 (42.9)
Preserved sensation	20 (95.2)	1 (4.8)
Signs of autonomic dysfunction		
Hypertension	2 (9.5)	19 (90.5)
Hypotension	3 (14.3)	18 (85.7)
Tachycardia	6 (28.6)	15 (71.4)
Bradycardia	1 (4.8)	20 (95.2)
Symmetry of weakness	19 (90.5)	2 (9.5)

Table 5 Investigations done to studied cases

Investigations	Yes [*n* (%)]	No [*n* (%)]	Notes [*n* (%)]
Lumbar puncture	6 (28.6)	15 (71.4)	
Free			3 (14.3)
Cytoalbuminologic dissociation			3 (14.3)
Nerve conduction studies	21 (100.0)	0	
Normal			2 (9.52)
Delayed F-wave			19 (90.48)

Table 6 Treatment given to studied cases

Treatment	Yes [*n* (%)]	No [*n* (%)]
IVIG	17 (81.0)	4 (19.0)
Plasmapheresis	3 (14.3)	18 (85.7)
Need for mechanical ventilation	6 (28.6)	15 (71.4)

IVIG, intravenous immunoglobulins.

of polio [11]. GBS has shown significant presence of preceding infection before its onset. In our study, history of preceding infection was found in 13 (61.9%) cases, with respiratory illness in most cases and diarrhea and nonspecific febrile illness in the remaining cases similar to Dhadke *et al.*[10] and Sadek *et al.* [12]. In our study, all cases presented with limb weakness at the onset of illness similar to Sadek *et al.*[12] and in correspondence to Dhadke *et al.* [10], who observed 13 of 40 cases in their

Table 7 Comparison of demographic and clinical characteristics of cases with two other studies

Characteristics	Dhadke *et al*. [10]	Kumar *et al*. [11]	Sadek *et al*. [12]	This study
Study population (*n*)	40	20	50	21
Study period	1 years 8 months	1 years 6 months	1 year	9 months
Age range	13 years to 40 years	16 months to 17 years	7 months to 11 years	18 months to 13 years
Male:female ratio	1.5:1	2.3:1	1.1:1	1.1:1
Preceding events prior to illness (%)	55	50	66	61.9
Clinical presentation (%)				
Limb weakness	100	100	100	100
Sensory affection	32.5	25	100	19
Cranial nerve affection	62.5	20	4	33.3
Autonomic dysfunction	None	25	26	28.6
Respiratory insufficiency requiring assisted ventilation	32.5	35	None	28.6
Albuminocytologic dissociation in CSF analysis (%)	65.3 (26 of 40)	40 (2 out of 5)	100 (50 out of 50)	50 (3 out of 6)
Recipients of IVIG (*n*)	14	16	50	17
Recipients of plasmapheresis (*n*)	4	1	None	3
Outcome (%)				
Complete recovery	75	82.4 (14 of 17)	78	80.95
Incomplete recovery	5	17.6	22	19.05

CSF, cerebrospinal fluid; IVIG, intravenous immunoglobulins.

study with sensory symptoms in the form of paresthesia, tingling, numbness, or pain. In our study, seven (33.3%) cases had cranial nerve involvement mostly in the form of chocking and absent gag reflex (bulbar affection) in correspondence to Dhadke *et al*. [10], who found 62.5% of their cases showing cranial nerve involvement mostly the facial nerve. In our study, six (28.6%) cases developed respiratory insufficiency and required assisted ventilation similar to Kumar *et al*. [11], who in their study found five (25%) cases suffering from respiratory insufficiency and requiring assisted ventilation. In our study, CSF analysis was done in six cases. Typical CSF finding of cytoalbuminologic dissociation was found in only three (14.3%) cases similar to Kumar *et al*. [11], who in their study performed CSF analysis on five cases. Typical CSF finding was found in only two cases (Table 7).

Conclusion

IVIG and supportive care are considered the key elements in the management of GBS in children. IVIG administration early in the course of the disease results in better recovery. Plasmapheresis may be an effective option in cases with poor response to IVIG. More studies are needed to be conducted on childhood GBS, on a wider base of cases, to gain more information about the disease and its treatment options and to achieve better results.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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