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Research Article

Diagnosis, Treatment and Prognosis of Guillain-Barré Syndrome in Children

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Abstract:

Aim: Guillain-Barre syndrome (GBS) is one of the reasons of acute polyneuropathy causing severe morbidity and mortality. In this study, it was aimed to identify the clinical findings, treatment modalities and factors affecting the prognosis of patients who were followed up and treated with GBS.

Material and Method: A total of 47 patients diagnosed and treated with GBS between 2006 and 2016 were evaluated in Child Health and Diseases Services. Parameters such as age, gender, presenting complaints, previous infection history, seasonal distribution, cranial nerve involvement, presence of autonomic symptoms, muscle weakness, need for respiratory support, electroneuromyography (ENMG) findings, length of stay in the intensive care unit and general wards, and treatment were investigated in the patients.

Results: Twenty nine (61,7%) of the patients were male, 18 (38,3%) were female and the mean age was 7,94±4,49 years. The complaints of the cases observed in the application; 44 patients (93,6%) had weakness and 3 (6,4%) patient had numbness in the feet. Twenty seven (57,4%) of the 47 patients had prior history of infection. Upper respiratory tract infections and gastrointestinal system infections were found in 14 (51,9%) and 12 (44,5%) cases respectively. Gender, presence of infection, type of infection, cranial nerve involvement, presence of autonomic symptoms, and subtype of GBS were not found to be prognostic. The duration of stay in intensive care unit was 9,43±7,81 days and the total length of stay in hospital was 19,32±14,62 days (p= 0,009). The need for respiratory support and intravenous immunoglobulin (IVIG) treatment were found to be effective factors on prognosis.

Conclusion: While the need for respiratory support in patients with GBS was a poor prognostic factor, it was observed that IVIG treatment alone could be associated with good prognosis. There is a need for extensive research to determine prognostic factors.

Keywords: guillain-barre syndrome; prognosis; children

Introduction

Guillain-Barré Syndrome (GBS) is an acute inflammatory polyneuropathy characterized by rapidly progressive, ascending, symmetrical weakness and areflexia. Its global incidence ranges from 0.6 to 2.4 per 100,000 individuals annually [1]. GBS can affect individuals of all ages and both sexes. Symptoms typically begin in the lower extremities and progress to the trunk and upper limbs within a few days. While progression is usually symmetrical, in rare cases, an asymmetrical onset may occur. Sensory loss, autonomic dysfunction, cranial nerve involvement, and neuropathic pain frequently accompany the clinical presentation.

GBS symptoms commonly appear following a viral or bacterial infection; more rarely, the condition may develop after vaccination [1]. There are both demyelinating and axonal variants of GBS, with differing prevalence across geographical regions. Acute inflammatory demyelinating polyneuropathy (AIDP) is the most common subtype in North America and Europe, accounting for approximately 90% of all cases [2]. In contrast, acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN) are more prevalent in Central and South America as well as in Asia [3].

The prognosis is generally more favorable in children than in adults. Several studies have investigated the factors influencing the prognosis of pediatric GBS [4-6]; however, data on childhood GBS cases remain limited. Identifying prognostic factors is crucial for optimizing treatment and follow-up strategies. The objective of this study was to evaluate the demographic and clinical characteristics, treatment approaches, and prognostic outcomes in pediatric GBS cases.

Materials and Methods

This retrospective study included 47 patients who were diagnosed with Guillain-Barré syndrome and received treatment and follow-up care between 2006 and 2016. Patient records were reviewed, and a standardized data collection form was completed for each individual. Ethical approval for the study was obtained from the institutional Ethics Committee.

Data collected included age, gender, presenting complaints, clinical findings at admission, history of preceding infection (including the type of infection), seasonal distribution, cranial nerve involvement, presence of autonomic symptoms, severity of muscle weakness, need for respiratory support, electroneuromyography (ENMG) findings, duration of stay in the intensive care unit and general wards, treatment protocols, and clinical status and examination findings at both discharge and two months post-discharge.

The examination findings of the patients at the time of admission and at the two-month follow-up after discharge were made according to the scale of Hughes et al [7]

0 - No findings

1 - Minor findings

2 - Walking without support

3 - Walking with support

4 - Bedridden

5 - Requiring respiratory support

6 - Death

Electroneuromyography was performed between 7 and 20 days after the onset of symptoms. Based on clinical and ENMG findings, patients were classified into three subtypes of GBS: acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), and acute motor and sensory axonal neuropathy (AMSAN). Autonomic symptoms were defined as the presence of high or low blood pressure, urinary retention, and cardiac dysrhythmias.

To evaluate prognostic factors, patients were divided into two groups (G1 and G2) based on their clinical findings at the follow-up visit conducted two months after discharge.

G1: 0 - No findings, 1 - Minor findings

G2: 2 - Walking without support, 3 - Walking with support, 4 – Bedridden, 5 - Requiring respiratory support, 6 – Death.

Treatment modalities were categorized as either intravenous immunoglobulin (IVIG) alone or a combination of IVIG and plasmapheresis. The length of stay in both the intensive care unit and general wards was also recorded.

Demographic and clinical data of all 47 patients were retrospectively analyzed using the statistical software package "SPSS for Windows, version 21.0". Descriptive statistics, t-tests, and chi-square tests were employed for data analysis. A p-value of <0.05 was considered statistically significant.

Results

This study included 47 patients diagnosed with Guillain-Barré syndrome. Of these, 29 were male (61.7%) and 18 were female (38.3%), with a mean age of 7.94 ± 4.49 years. Thirty-two patients (68.1%) were under 10 years of age, and 15 (31.9%) were over 10 years of age. Among those under ten years, 20 patients (62.5%) were classified in Group 1 (G1) and 12 (37.5%) in Group 2 (G2). Of the patients over 10 years of age, 10 (66.7%) were in G1 and 5 (33.3%) in G2.

A history of preceding infection was reported in 57.4% of the patients, the majority of which were upper respiratory tract infections (URTIs). The disease most frequently occurred in the spring.

Initial clinical evaluations showed that 44 patients (93.6%) presented with limb weakness, while 3 (6.4%) reported numbness. Muscle weakness was symmetrical in all patients. In 16 patients (34.0%), only the lower extremities were affected, while 31 patients (66.0%) exhibited involvement of both lower and upper extremities. Cranial nerve involvement was observed in 10 patients (21.3%), and autonomic symptoms were noted in 8 patients (17.0%). Among those with autonomic involvement, 6 patients (75%) exhibited blood pressure fluctuations. One patient had both blood pressure changes and urinary retention, while 3 patients (37.5%) had only urinary retention. Respiratory support was required in 13 patients (27.7%).

Among the 47 patients, 18 (38.3%) were diagnosed with AIDP, 18 (38.3%) with AMAN, and 11 (23.4%) with AMSAN with electrophysiological findings.

Regarding treatment, 25 patients (53.2%) received intravenous immunoglobulin (IVIG) alone, while 22 patients (46.8%) received a combination of IVIG and plasmapheresis. General characteristics of the patients are presented in Table 1.

The mean length of stay in the intensive care unit was 9.43 ± 7.81 days, and the mean total hospital stay was 19.32 ± 14.62 days.

For prognostic evaluation, patients were divided into two groups based on clinical findings at the two-month follow-up visit after discharge. Thirty patients (63.8%) were classified as G1 and 17 patients (36.2%) as G2. When evaluating prognostic factors, no statistically significant relationship was found between prognosis and variables such as gender, age, history of prior infection, type of infection, seasonal distribution, cranial nerve involvement, or the presence of autonomic symptoms. However, the need for respiratory support and the type of treatment administered were found to have a statistically significant impact on prognosis. Prognostic factors are summarized in Table 2.

Discussion

Guillain-Barré Syndrome is a common, immune-mediated, acute inflammatory demyelinating polyradiculopathy affecting the peripheral nerves. In the pediatric population, GBS is more prevalent in boys than in girls, with the incidence approximately 1.5 times higher in boys (8). In a study by Korinthenberg et al. (9), the female-to-male ratio was 1:1.27, and the age range was between 11 months and 17.7 years. In our study, 29 patients (61.7%) were male and 18 (38.3%) were female, with a female-to-male ratio of 1:1.6 and a mean age of 7.94 ± 4.49 years, which aligns with the existing literature.

Approximately two-thirds of GBS patients report a preceding infection. Upper respiratory tract infections (URTIs) are the most common, followed by gastroenteritis caused by *Campylobacter jejuni* (10). In a study by Varkal et al. (11), 32 out of 40 patients (80%) had a history of

preceding infection, with 21 cases (65.6%) being URTIs and 11 (34.4%) gastroenteritis. In our study, 57.4% of patients had a preceding infection: 14 patients (51.9%) had URTIs, 12 (44.5%) had gastrointestinal infections, and 1 patient (3.6%) had a history of vaccination Additionally, most patients presented in the spring.

In typical GBS, rapidly progressive bilateral weakness is the hallmark symptom. The weakness classically begins in the distal lower extremities and ascends proximally. However, it may sometimes begin proximally or in the upper extremities. Akbayram et al. (13) reported that muscle weakness was the initial symptom in 34 of 36 patients (94.4%), with muscle pain reported in 8, dysarthria in 3, and numbness in 2. Similarly, in our study, 44 patients (93.6%) presented with limb weakness and 3 (6.4%) with numbness in the feet.

GBS is classified into axonal and demyelinating subtypes based on clinical and electrophysiological findings. AIDP is the most common clinical form. Previous studies have suggested differences in clinical features and prognosis among subtypes [14]. Akbayram et al. [13] reported that 25 of 36 patients (69.4%) had AIDP, 10 (27.8%) had AMAN, and 1 (2.8%) had AMSAN, with no significant difference in prognosis between subtypes. Nagasawa et al [15] found that the AIDP subtype was more commonly associated with respiratory tract infections and showed greater cranial and sensory nerve involvement, whereas the AMAN subtype was more commonly linked to gastroenteritis and had purely motor involvement. Although both subtypes were associated with good outcomes, recovery was slower in AMAN patients. In our study, 18 patients (38.3%) had AIDP, 18 (38.3%) had AMAN, and 11 (23.4%) had AMSAN. No significant association between subtype and prognosis was observed. Despite reports suggesting a poorer prognosis in axonal forms, we did not find a difference in our cohort, possibly due to the limited sample size.

Cranial nerve involvement is also observed in GBS patients. While 9th and 10th cranial nerve involvement is more common in younger children, facial nerve involvement is frequently reported in older children [16]. Although the association between cranial nerve involvement and prognosis remains unclear, some studies have suggested that these patients may be at higher risk of respiratory muscle involvement, requiring intubation, and may have worse outcomes [16]. Autonomic symptoms such as tachycardia, hypotension, and sinus arrhythmia are frequently observed, particularly in the AIDP and AMSAN subtypes and in patients with quadriplegia and respiratory failure [17]. Several studies have demonstrated an association between autonomic symptoms and increased need for respiratory support [18,19]. DiMario et al [20] reported autonomic dysfunction in 24 of 26 patients (92.3%) and established a correlation with disease severity. In our study, 10 patients (21.3%) had cranial nerve involvement, and 8 (17.0%) exhibited autonomic symptoms. However, neither cranial nerve involvement nor autonomic dysfunction was found to be significantly associated with prognosis.

Respiratory failure occurs in 20–30% of GBS cases and is associated with the rapidity of disease progression. The prognosis is generally worse in children requiring respiratory support. Mortality in ventilated patients has been reported to be between 15% and 30% [21]. In a study by Kalita et al [22] involving 328 GBS patients, including 85 under 15 years of age, 43 required ventilatory support. Of these, 26 had good outcomes, and 17 had poor outcomes, with no significant difference in prognosis between the groups. In contrast, Fletcher et al [23]., in a study of 114 cases, found that 60 patients required respiratory support and that 81% of patients with poor prognosis had received mechanical ventilation. They concluded that the need for respiratory support was associated with worse outcomes. In our study, 13 patients (27.7%) required respiratory support, and these patients had a slower recovery. We also found that respiratory support was significantly associated with poor prognosis.

Both intravenous immunoglobulin (IVIG) and plasmapheresis have been proven effective in GBS treatment [3,12]. IVIG is generally preferred in children due to its ease of administration and lower risk of complications. Van der Meché et al [24], in a randomized trial involving 147 patients (74 treated with IVIG and 73 with plasmapheresis), found no significant difference in prognosis between the two treatments. In our study, 25 patients (53.2%) were treated with IVIG alone, while 22 (46.8%) received both IVIG and plasmapheresis. At the two-month follow-up, 22 of the 25 patients treated with IVIG alone were in Group 1 (good prognosis), whereas only 8 of the 22 patients who received combination therapy were in Group 1. Most of the patients in the combination group were classified as Group 2. These findings indicate that patients treated with IVIG alone had a significantly better prognosis than those treated with IVIG plus plasmapheresis.

In pediatric GBS, the prognosis is generally more favorable than in adults. However, the prognostic factors remain incompletely understood. Various studies have explored clinical and electrophysiological predictors. In our study, factors such as age, gender, antecedent infection, cranial nerve involvement, and autonomic symptoms were not significantly associated with prognosis. However, the need for respiratory support and the type of treatment administered were significant prognostic indicators. We observed that the prognosis was worse in patients who required respiratory support and in those treated with a combination of IVIG and plasmapheresis.

In conclusion, our study suggests that in pediatric GBS, gender, age, preceding infection, seasonality, cranial nerve involvement, and autonomic symptoms do not significantly affect prognosis. In contrast, the need for respiratory support and the treatment method were significant predictors of outcome.

Conflict of interest: Authors declare that there is no conflict of interest between the authors of the article**.**

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Data availability: Data are available on reasonable request.

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