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Original Article

Guillain–Barré Syndrome in the United Arab Emirates: A Sixteen-Year Experience of a Single Center

Salim Yaghi[1], Waleed S. Beshyah[1], Anas S. Beshyah[1], Aliasgar Lokhandwala[2], Salem A. Beshyah[3]

[1]Internal Medicine Residency Program, Institute of Education and Divisions of [2]Internal Medicine and [3]Endocrinology, Institute of Medicine, Sheikh Khalifa Medical City, Abu Dhabi, United Arab Emirates

Abstract

Background: Guillain–Barré Syndrome (GBS) is the most common cause of acute flaccid paralysis. We describe clinical characteristics and outcomes of GBS in a region where data are hitherto limited. **Patients and Methods:** This retrospective study was conducted at Sheikh Khalifa Medical City, Abu Dhabi, United Arab Emirates. All GBS patients identified between 2000 and 2015 were documented and summarized using descriptive statistics. **Results**: Fifty-three patients were identified. GBS was 3.1 fold more common in men; median age was 33 (range: 16–79). 47.2% of the patients were previously healthy. Upper respiratory tract infections occurred in 37.7% and gastroenteritis in 18.9%. The most common clinical presentation was bilateral upper and lower limb weakness (47.2%). Vital capacity was low in 63.16% of patients. Classical albuminocytologic dissociation was seen in 19.5% of patients who had a lumbar puncture done. Nerve conduction studies were carried out in 56.6% of cases, and the most commonly observed abnormality was mixed axonal neuropathy. Nearly 60.38% of patients had adverse outcomes. Eight patients (25%) required admission to the intensive care of whom six required mechanical ventilated. Twenty-Four patients (75%) were discharged with residual motor weakness, and five of whom required further rehabilitation. Poor respiratory status on arrival significantly predicted poor outcome. **Conclusions:** Our results are generally similar to international trends in GBS. Respiratory dysfunction and decreased vital capacity were the only poor prognostic factors identified. Nerve conduction findings were not associated with clinical or prognostic values.

Keywords: Clinical pattern, epidemiology, Guillain–Barré Syndrome, incidence, neuropathy, United Arab Emirates

Introduction

Guillain–Barré Syndrome (GBS) is a form of acute inflammatory peripheral polyneuropathy. It is currently the most common cause of acute flaccid paralysis worldwide.[1] In the vast majority of patients, the disease presents with progressive, ascending motor paralysis and areflexia. GBS has an incidence of 1–2 cases per 100,000 a year worldwide with males being affected more often than females.[1]

In the acute form of the illness, patients usually present with progressive ascending paresis or paralysis that is typically symmetrical and begins distally and extends to affect the more proximal muscles. The clinical manifestation of GBS is usually preceded by a febrile illness, most commonly an upper respiratory or gastrointestinal tract infection. However, in some patients, the trigger can be a noninfectious stressor such as surgery, trauma or pregnancy. Other key features of GBS include the involvement of facial and bulbar muscles, autonomic disturbances, and of great importance is the effect on the muscles of respiration which can be severe enough to cause respiratory dysfunction or failure leading to the need for intubation and mechanical ventilation or sudden death.[1-3]

Despite the advances in GBS diagnostic investigations, such as nerve conduction studies (NCSs), and treatment modalities, such as intravenous immunoglobulins or plasmapheresis, mortality rates associated with this condition remain as high as 5%–10%.[3] In addition, even in patients who receive treatment, approximately 20%–25%

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Address for correspondence: Dr. Anas S. Beshyah, Internal Medicine Residency Program, Institute of Education, Sheikh Khalifa Medical City, P. O. Box 59472, Abu Dhabi, UAE.
E-mail: anas.beshyah@gmail.com

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are left with significant morbidity in the form of persistent disability.[3]

Clinical studies analyzing the demographics, trends, and outcomes of patients affected with the condition especially in the Middle East and Gulf region are limited.[4] This study aims to add potentially valuable information to the limited pool of data concerning GBS clinical variables, treatment, and outcomes within our region. In this study, we try to ascertain the demography, clinical trends, and presentation of patients with GBS in a tertiary care setting (Sheikh Khalifa Medical City (SKMC), Abu Dhabi, United Arab Emirates (UAE)) over the course of 16 years (2000–2015).

Patients and Methods

This retrospective study looked at GBS data collected in a tertiary care center in the UAE (SKMC) over a period of 16 years. Approval of the study was obtained from the SKMC Institutional Review Board. Hospital electronic databases were searched for patients admitted with suspected or confirmed Guillain–Barré Syndrome and charts were retrieved and reviewed. A total of 53 patients fulfilled the inclusion criteria and were admitted in the 16-year window between the year 2000 and 2015 (inclusive). A predefined dataset was extracted and tabulated in an electronic spreadsheet. Patient demographic data, clinical presentation and symptoms on admission, known baseline medical comorbid conditions, history of recent preceding illness (e.g., diarrheal disease or upper respiratory tract infection) were extracted. The details of neurological examination findings with particular attention to cranial nerve involvement were noted. Recorded respiratory dysfunction (based on Vital Capacity assessments), arterial blood gas results, the need for intubation and mechanical ventilation and hospital length of stay were captured. Diagnosis of GBS according to one or more of the following: clinical criteria, lumbar puncture (LP) findings, and NCSs. Pediatric patients (younger than 15-years-old) and those with other causes of flaccid paralysis other than GBS, as evident by brain imagining (computed tomography [CT] or magnetic resonance imaging [MRI]), were excluded from the study. The results of diagnostic studies were reviewed including LP findings (e.g., biochemistry, cells counts, and presence of antibodies), NCSs, and brain imaging (CT and/or MRI). The patient's treatment regimens including, but not limited to, the use of intravenous immunoglobulins, plasmapheresis, and other management options, were documented. Clinical outcome was based on physical status at the time of discharge (i.e., patients were grossly divided into those with or without residual weakness and patients with worsening weakness requiring rehabilitation).

Results

Demographics and premorbid status

A total of 53 patients of mixed ethnicity were included [Table 1]. The majority of the patients were males (75.5%). Male-to-female ratio was 3.1:1.0. The mean age at onset was 36 (33) years. The median (range) was 33 (16–79) years. 28 (52.8%) of the patients were known to have preexisting comorbidities, and 25 patients (47.2%) were previously healthy. Twenty patients (37.7%) of patients had a recent upper respiratory tract infection, probably as the precipitating cause of GBS. Ten cases (18.9%) had gastroenteritis before presentation. In 9 patients (17.0%), the trigger for GBS was attributed to a indetermined febrile illness, and the remaining 14 cases (26.4%) had no documented identifying precipitating factor.

Table 1: Demographic, gender, prior health, and ethnicity data

Variables	Results
Age: Mean; median (minimum-maximum)	36; 33 (16-79)
Gender (males/females) *n* (%)	40:13 (75.5%:24.5%)
Ethnicity, *n* (%)	
Africa	6 (12.8)
Asia	16 (34.0)
Europe	3 (6.4)
Middle East	27 (57.5)
North America	1 (2.1)
Premorbid state, *n* (%)	
Healthy	35 (66.0)
Preexisting comorbidities	18 (34.0)

Neurological deficits

The most frequent presentation was bilateral, combined, upper and lower limb weakness in 25 cases (47.2%). Bilateral lower limb weakness without upper limb involvement was seen in 21 patients (39.6%). No weakness was detectable in 4 patients (7.5%). Two patients (3.8%) presented with unilateral limb weakness and in a single case, only upper limb weakness occurred with sparing of the lower extremities. One-third of the patients (34%) had peripheral neuropathy in addition to the motor deficit. Cerebellar symptoms (vertigo, dizziness, or diplopia) were evident in 9 patients (17.0%). Detailed neurological examination including muscle tone, deep tendon reflexes, and cranial nerve affection extracted from charts [Table 2]. Hypotonia was seen in 14 patients (26.4%). Upper limb and/or lower limb hypo or areflexia was seen in 33 (62.3%) and 44 cases (83.02%), respectively. Cranial nerve involvement was seen in 20 patients, 11 of whom had isolated facial nerve palsy.

Diagnostic investigations

An LP was performed in 41 patients (77.4%). In 28 cases, cerebrospinal fluid (CSF) protein levels were above 0.4 G/L. The mean was 1.28 g/l (range 0.16–20 g/L). Cytological evaluation of the CSF showed <5 white blood cells (WBCs) in 30 patients. The mean CSF WBC count was four cells (range 0–50 cells). CSF immunoglobulins were measured in 12 of the 41 who had had an LP; 7 had elevated immunoglobulin G (IgG) levels, 4 had results within the normal range, and in one case the IgG value was low. NCSs were carried out in 30 patients. Mixed axonal neuropathy was found in 12 patients and motor axonal

Table 2: The frequency of abnormal neurological examination findings

Neurological examination findings	Patients' data: Number (%)	
Tone		
Normal	39 (73.6)	
Hypotonia	14 (26.4)	
Hypertonia	0	
	Upper limb (%)	**Lower limb (%)**
Power		
0	1 (1.9)	0
1	2 (3.8)	2 (3.8)
2	5 (9.4)	15 (28.3)
3	17 (32.1)	17 (32.1)
4	12 (22.6)	11 (20.8)
5	16 (30.2)	8 (15.1)
Deep tendon reflexes		
Absent/hyporeflexia	33 (62.3)	46 (86.8)
Normal	20 (37.7)	6 (11.3)
Hyperreflexia	0	1 (1.9)

neuropathy in 10. Brain imaging by CT, MRI brain, or both, were requested in 41 patients. Imaging studies reported no abnormalities in 35 patients.

Management and outcomes

Twenty patients were admitted to a general medical ward; 13 patients required admission to monitored beds and eight patients were admitted to the Intensive Care Unit. All patients received intravenous immunoglobulin (IVIG) treatment during their hospitalization. A small group, five patients, was also given additional treatment modalities (three plasmapheresis, 2 IV steroids while receiving IVIG therapy). IgA levels were measured in 11 patients before initiating IVIG treatment of whom only one had low IgA level. There was no record of any anaphylactic reactions to therapy. Respiratory distress necessitating intubation and mechanical ventilation occurred in 6 patients (11.3%). However, vital capacity (VC) was documented in 19 only. Of these, 12 and seven patients had VC less and less 1.2 L respectively. Of the six patients that assisted ventilation 3 had VC values, greater and 3 had VC <1.2 L. In the remaining cases who needed assisted ventilation, two required intubation for clinically evident respiratory failure and one patient after arterial blood gas analysis revealed respiratory acidosis. The most common ethnicities were Middle Eastern (50.9%) and East Asians (30.2%). Adverse outcomes were observed in 56.2% and 66.7% of the two groups respectively. No statistically significant correlation could be found between the ethnicity and adverse outcomes. On discharge, 21 patients had no residual weakness, 27 patients had some residual, and 5 patients had worsening weakness and required lengthy physical rehabilitation. The length of stay ranging from 4 to 235 days with a mean length of stay of 30.6 (median 13) days. Adverse outcomes were observed in more than half of our patients (60.38%). Adverse outcome had statistically significant associations only with respiratory dysfunction on arrival ($\chi^2 = 4.101$; df = 1; $P = 0.043$).

Discussion

In the present study, we aimed to collect, analyze, and present data on GBS from a new geographical location that has not been examined previously. Understandably, this is a retrospective study, meaning it looked backward and examined exposures to suspected risk or protection factors concerning an outcome that is established at the beginning of the study. Many valuable case–control studies published in the literature were retrospective investigations. They remain a readily available source of literature in some regions of the world. The main conclusion was that the clinical characteristics of GBS in the UAE are similar to GBS elsewhere. Overall, the study did not provide novel findings, and it attempted to give some information about the clinical characteristics of GBS in the UAE to complete the regional and global epidemiological picture.

The most important source of bias in the present study is the small sample size of patients. This is predictable on two counts; first, the study being was conducted in a single center, albeit a referral center, and second, GBS being a rare disease as inferred from some of the previous studies. The authors are conscious of the concern of the small sample size. In particular, being careful with any temptation to compare the between the outcomes of different treatments. Notwithstanding, we realize this being potentially a fascinating aspect.

Our patients had antecedent infection in a 57% of cases which is lower than previously reported 75%.[2,3] The characteristic clinical presentation of GBS is that of bilateral, ascending lower extremity weakness was the most common presentation in our patients, with 87% of patients exhibiting bilateral lower limb weakness (47% of these patients also had bilateral upper limb weakness in addition to their lower limb symptoms). It is noteworthy that in 80% of cases paresthesia affecting both the hands and feet can accompany the lower limb weakness, as mentioned in relevant literature. In such cases, the clinical examination of the patients fails to demonstrate any sensory abnormality, or if present it is limited.[5-7] Eighteen (34%) of the study population complained of subjective sensory neuropathy, however, on examination, no more than 10 had sensory deficit. The empirical diagnosis of GBS heavily relies on the clinical presentation and identification of the typical pattern of symmetrical, progressive ascending muscle weakness that can range from mild weakness to complete paralysis of motor, respiratory, facial, or bulbar muscles, associated with hyporeflexia or areflexia. A definitive diagnosis is generally made when the CSF and/or nerve conduction study results corroborate the clinical suspicion.[6,7]

Albuminocytologic dissociation is typically found in patients with GBS. However, it is important to note that CSF protein concentrations are normal in up to 50% of cases during the 1st week of illness and this increases to 75% in the 3rd week.[1,3,7,8]

The majority of our study population (41 patients, 77.35%) underwent LP as part of their work up. Cerebrospinal fluid analysis revealed elevated protein concentrations, defined as values >0.4 g/L, in 28 patients (68.29% of patients who had a LP) while 30 patients (73.17%) had low CSF white blood cell counts, defined as <5 cells per high-power field. Eight patients out of 41 (19.5%) had albuminocytologic dissociation seen in CSF analysis.

Respiratory dysfunction secondary to GBS can be a medical emergency and is generally observed in up to 25%–30% of GBS patients.[1-3,9] Respiratory insufficiency requiring intubation and mechanical ventilation was seen in 6 patients (11%). Respiratory function is monitored using VC along with the patient's vital signs (including oxygen oximetry) every 6–12 h with continuous cardiac monitoring if the patient is clinically stable.[10] In patients with more progressive and severe disease, and those deemed clinically unstable, this should be performed every 2–4 h.[10] Of the 53 patients included in the study, only 19 had VC recordings documented.

NCSs can be a useful means of confirming the diagnosis of GBS and may also be used to further divide patients into either demyelinating or axonal subtypes.[11-13] Worldwide, the most common subtype of GBS is autoimmune demyelinating polyneuropathy which comprises more than 85% of all GBS cases.[11] The remaining 5%–10% of cases display an axonal neuropathy pattern of disease and can further be divided into either axonal motor neuropathy or axonal sensorimotor neuropathy.[12]. The prevalence of the different subtypes varies slightly in different regions.[11] It is worth noting that, when performed during the late course of the disease, the nerve conduction study may lead to a more substantial portion of patients being incorrectly labeled as having the axonal variant of the disease.[13] This phenomenon is thought to be due to secondary axonal degeneration of patients with demyelinating GBS.[13] The most commonly observed GBS subtype in our study was mixed axonal neuropathy which was seen in over a third of patients (12 of the 31 patients, 39%). The second most common abnormal conduction pattern was motor axonal neuropathy (10 patients, 32% of our study population). In our study population, the demyelinating subtype was found to be the least common and was seen in only three patients (9% of the total). It is possible that this figure may be underestimating the prevalence of demyelinating GBS in the UAE as NCSs were not performed in 22 patients, of which 20 (91%) did not experience any clinical deterioration, which is characteristic of the demyelinating subtype. Furthermore, NCSs may also overestimate the number of axonal cases when performed late in the course of the disease when secondary axonal degeneration sets in as part of the natural course of the disease. This may explain the unexpectedly large portion of patients in our study with axonal neuropathy on NCS.

Treatment of GBS includes general supportive measures and specific management.[1,8,11] For the specific therapy, either IVIG or plasma exchange is recommended as a treatment option within the first 2–4 weeks after the onset of the disease.[14-18] Neither treatment modality was superior and that there was no added benefit when combining IVIG and plasmapheresis treatment.[14,15] There is no role for glucocorticoids therapy.[16] All the patients in our study received a course of IV immunoglobulin, besides three received concurrent plasmapheresis therapy, and two patients received IV steroid treatment. Screening for IgA deficiency in patients before the initiation of IVIG treatment as patients with this deficiency may be predisposed to develop anaphylactic reactions when treatment is commenced. It is believed that the development of anti-IgA antibodies causes this in these at-risk patients and so when IgA containing IVIG solutions are administered, severe allergic reaction may occur.[17,18] Only one patient was documented to have low levels of IgA with no adverse reaction after treatment.

Long-term outcomes vary among patients with GBS but, with appropriate treatment, 80%–84% of patients can walk independently within 6 months of diagnosis. The majority of patients (60%) regain full motor power. Despite advances in treatment and high-level hospital care, 5%–10% of affected patients have complicated clinical courses, complications during their hospital stay, and require prolonged duration of mechanical ventilation. Five percent of patients die despite intensive care.[19] Adverse outcomes were observed in more than half of our patients (60.38%). Adverse outcome had statistically significant associations with respiratory dysfunction on arrival which reflects the acute severity and need for ventilation, etc., Notwithstanding, lack of long-term follow-up to identify patients who regain their muscular power following discharge from hospital could explain the relatively high percentage of adverse outcomes, as the natural course of illness may take up to 4 weeks to show full recovery.[19,20]

Perhaps, the main limitations of this study are inherent in its retrospective design, single-center site pool, small number of patients identified, and the lack of follow-up for a more extended period compatible with the recognized natural history of GBS. Lack of a unified management protocol resulting from the fact that thoughts, practices, and physicians changed over the years must be mentioned too.

Conclusions

Although small in size, the present study provided new insight into the clinical trends of GBS in the UAE. The trends observed are similar to international data about the disease. A male predominance, premorbid infections were common, and bilateral LL weakness was the most frequent pattern of presentation. National or even transnational prospective studies are needed to ascertain the epidemiology and outcomes concerning known or novel risk factors in the Gulf or the Middle East.

Authors' contributions

All authors contributed to the conception and planning of the study. SY, WSB, ASB performed data collection, and analysis

and drafted the manuscript. AA and SAB provided expertise and guidance and revised the data analysis and manuscript. All authors approval of the paper in its final version.

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Conflicts of interest

There are no conflicts of interest. SAB is the father of WSB and ASB.

Compliance with ethical principles

Ethical approval was granted by the Institutional Review Board of SKMC (reference number RS-406). All patients admitted to our institution sign a general consent for their data to be used in research anonymously. Neither specific consent is feasible nor required for retrospective chart reviews and audit type of exercise.

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Reviewers:	Editors:
Khadija Ahmed Hafidh (Dubai, UAE) Abdulwahab El Barsha (Benghazi, Libya)	Elmahdi Elkhammas (Columbus, Ohio, USA)