The Guillain-Barré Syndrome at Two Hospitals in Trinidad, West Indies
A Review of 26 Patients.
A Esack, S Teelucksingh

ABSTRACT
Over a four-year period, 26 consecutive patients with Guillain-Barré syndrome (GBS) were seen. Their ages ranged from 18 months to 68 years. Fifteen were male and 11 female. The crude annual incidence was estimated to be 1.5 per 100,000 population. East Indians made up the majority of the patients. An antecedent infection was reported in 65% of patients. Significant pain was present in half of the cohort. F-wave abnormalities were the commonest electrophysiological disturbance. Twenty-nine per cent of patients required ventilation. Intravenous immunoglobulin (IVIG) treatment was beneficial in 88% of patients. Eighty-four per cent made a complete or near complete recovery. One patient died.

Keywords: Guillain-Barré Syndrome, Trinidad

INTRODUCTION
The Guillain-Barré syndrome (GBS) is an acute or subacute polyradiculoneuropathy. The disease affects 1–2: 100,000 population annually (1). The main pathological process is demyelination of the peripheral nerves, although more severe cases may be associated with predominant axonal changes (2). Both cellular and humoral factors are involved in the pathogenesis (3). The illness is frequently preceded by an infection, with Campylobacter jejuni being implicated in up to 40% of patients (4). Pain is a relatively common and often neglected part of the disease (5).

Nerve conduction studies/electromyography (NCS/EMG) typically shows features consistent with demyelination and/or axonal damage (6). Small compound muscle action potentials (CMAPs) are related to a worst prognosis (7).

Plasma exchange (PE) leads to a more rapid improvement than supportive care alone (8) and intravenous immunoglobulin therapy (IVIG) has been shown to be at least as effective as PE (9).

There are some geographical differences in several features of GBS. For instance, in Northern China, a pure axonal form of the illness exists (10). The purpose of the study was to examine various aspects of the illness as it

Palabras claves: Síndrome de Guillain-Barré, Trinidad

RESUMEN
Por un periodo de cuatro años, se atendieron 26 pacientes consecutivos con el síndrome de Guillain-Barré (GBS). Sus edades fluctuaban de 18 meses a 68 años. Quince eran varones y 11 hembras. Se calculó que la incidencia anual bruta era 1.5 por 100,000 población. La mayor parte de los pacientes eran indo-orientales. El 65% de pacientes reportó antecedentes de infección. La mitad de la cohorte presentaba dolor significativo. El trastorno electrofisiológico más común fue las anormalidades de la onda F. Veintinueve por ciento de los pacientes necesitaron ventilación. El tratamiento de inmunoglobulina intravenosa (IVIG) fue beneficioso en 88% de los pacientes. Ochenta y cuatro por ciento tuvo una recuperación completa o casi completa. Un paciente murió.

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From: The Eric Williams Medical Sciences Complex, Champs Fleurs, Mt Hope, Trinidad and Tobago, West Indies

Correspondence: Dr A Esack, The Eric Williams Medical Sciences Complex, Mt Hope, Trinidad and Tobago, West Indies, E-mail: neuroservices@gmail.com

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occurred in a Southern Caribbean island, Trinidad, and to draw comparisons with GBS in other countries.

PATIENTS AND METHODS
A retrospective analysis of 26 consecutive patients with GBS presenting to two hospitals was undertaken. The patients all had typical clinical features of the illness with rapidly progressive weakness affecting the limbs. In addition, some patients had involvement of the bulbar, facial and respiratory muscles. The deep tendon reflexes were either depressed or absent and abnormal sensory signs were largely lacking. The GBS subtypes included acute inflammatory demyelinating neuropathy (AIDP) and acute motor-sensory axonal neuropathy (AMSAN). All patients fulfilled the internationally accepted criteria for the diagnosis of GBS (11) [Appendix 1]. The study examined patients over a four-year period. Each patient was followed for at least one year. The following data were obtained: age, gender, ethnicity, a history of preceding infection (as judged by fever, diarrhoea or upper respiratory symptoms), the time interval between onset of the illness (sensory complaints or weakness) and presentation, presence of pain, functional score (FS) [Appendix 2] at presentation and one month after treatment plus NCS/EMG. Blood tests included Human T-Cell Lymphotrophic Virus-1 (HTLV-1) and Human Immuno-deficiency Virus (HIV) status (consent obtained), plus plasma creatinine phosphokinase (CPK) levels. The cerebrospinal fluid results were also noted.

NCS/EMGs (Cornblath’s electro clinical criteria (6) and CSF examination were both done on day 1 of admission using standard techniques. The dosage of intravenous immunoglobulin (IVIG) used was 0.4 g/kg/day for five consecutive days. Patients with an FS score of 3 and above were treated with IVIG.

RESULTS
Ages ranged from 18 months to 68 years (average 31 years). There were six patients under the age of 14 years. Fifteen patients (57%) were male and eleven (43%) female. Sixteen patients (61%) were of East Indian origin, 6 patients (23%) African, 3(11%) of mixed descent and one (5%) Caucasian.

Eight patients (30%) had a FS of 5, thirteen (50%), a FS of 4, three (11%) a FS of 3 and two patients (9%) a FS of 2. Seventeen patients (65%) reported an antecedent infection one to four weeks prior to the onset of their illness. Four (25%) had diarrhoea and six (35%) upper respiratory tract infections. Six patients (35%) had a non-specific febrile illness. One patient (5%) had dengue fever and one was found to be HTLV-1 positive.

Patients with a FS of 5 had an average time interval of 4.8 days (range 1–14 days). For a FS of less than 5, the interval was 6.7 days (range 1–21 days). Fourteen out of the twenty-six patients had significant pain (requiring analgesia). Back pain was present in six patients (23%), muscle pain in six patients (23%) and neck pain in three patients (12%).

The plasma CPK was elevated in four out of seven patients (57%) with pain and in one out of five patients (20%) without pain. Predominant peripheral demyelination was present in twenty-two patients (84%). Predominant axonopathy (sensory motor) was seen in three patients (11%). In one patient (5%), the NCS/EMG was normal. Of the twenty-two patients with predominant demyelination, F wave abnormalities (absent, prolonged or small) were found in 20 (90%), nerve conduction velocity slowing occurred in 4 (15%) and indeterminate in two (9%). The distal motor latency was increased in 16 (72%) out of the twenty-two patients, conduction block was seen in thirteen (59%) and dispersion in ten (45%) with ten patients (38%) having both F wave abnormalities and prolonged distal motor latencies but with normal nerve conduction velocities.

For patients with a FS of 5, the average CMAP size (mV) for the right median nerve (wrist) was 2.6 (0.1–11.0) and 0.14 (absent–0.5) for the right common peroneal nerve (ankle). When the FS was less than 5, the respective CMAP sizes were 4.3 (1.0–10.0) and 2.9 (0.1–7.5). Eight patients (29%) required ventilation. The average number of days on the ventilator was fifteen (7–42). All ventilated patients received IVIG. Of the eighteen patients treated with IVIG, sixteen (88%) improved by FS of 1 or more at one month. In the non IVIG group four out of eight patients (50%) showed a similar improvement. In the IVIG treated group, the average time to independent walking was 10 weeks (range 1 week to 11 months) and 12 weeks (range 0 to 1 year) in the non-IVIG group. The CSF was examined in eleven of the patients. In seven of these (63%), the CSF protein was elevated.

DISCUSSION
In Trinidad, roughly half of the population is of East Indian origin and half African. In this study group, Guillain-Barré syndrome was three times more common in the East Indians when compared to the Africans (ratio 2.7:1.0). Moreover, in other African predominant southern Caribbean islands such as Tobago and Grenada the incidence of the illness is extremely low (personal communication).

Comparisons must be treated guardedly since this was not a community based study and only data from two hospitals were reviewed. This makes the crude incidence of the disease of 1.5/100 000 population. This may or may not be fairly representative but this figure is similar to published incidences of GBS in North America (12) and the United Kingdom (13). The reported percentage of GBS patients with an antecedent infection is very much the same in this study group as in other countries (14). In one patient, dengue fever preceded the onset of GBS by two weeks (15). The HTLV-1 was positive in one patient. This could well be a chance occurrence since the overall prevalence of HTLV-1 positivity is three per cent in the Caribbean region. However, there is one case report of an
HTLV-1 associated polyradiculoneuropathy in a patient with adult T-cell leukaemia (16).

Although not statistically significant \( p > 0.05 \), patients who were more severely affected tended to have a shorter time interval from onset of illness to its nadir. This relationship has been previously reported (17). In this study, half of the patients complained of significant pain, primarily in the back, neck and muscles. Others have reported similar findings (5). The raised plasma CPK in four out of seven individuals with pain suggests that the pain may at least be due in part to changes in the muscle secondary to neurogenic damage. NCS/EMG showed that eighty-five per cent of the patients had predominant demyelinating disease and fifteen per cent had predominant axonal change. Slowing in the peripheral nerves was only present in a small number of individuals, the most common electrophysiological abnormality being a disturbance of the F wave. This observation is reported in other studies (17). In addition, nine patients had simultaneous F wave abnormalities and prolongation of the distal motor latencies, suggesting that at least early on in the disease, demyelination occurs both proximally and distally. Both the right median \( (p < 0.05) \) and right common peroneal \( (p < 0.05) \) CMAPs were smaller in patients with FS 5. The right common peroneal CMAP was particularly small in patients requiring ventilation. Correlation with CMAP size and prognosis in GBS has been cited by other workers (17). The pure acute motor axonal form of GBS (AMAN) was not seen in the present study. There was a favourable response to IVIG. Eighty-eight per cent improved by a FS of one or more at one month. Only fifty per cent showed a similar improvement in the non-IVIG group. This result is statistically significant \( p < 0.05 \). Van der Meche reported a fifty-three per cent similar improvement in his patients (9). The average time to independent walking was better in the IVIG group. This result is particularly significant considering that the IVIG group of patients was more severely affected. The treatment was well tolerated with no complications. Two patients given IVIG relapsed two to four weeks after treatment and a third relapsed two years after the initial illness. There is a small increased risk of relapse in GBS patients receiving IVIG (18).

To date, twenty-five of the twenty-six patients are ambulant. One ventilated patient died as a result of cardiac decompensation probably secondary to autonomic failure. Four out of the twenty-five ambulant patients are unable to run. The disability rate for patients with GBS in this small study in Trinidad is comparable to that reported by Winer and Hughes (19).

In summary, GBS, in this study from Trinidad, shares many similarities to GBS in other countries, including incidence, electrophysiology and disability rates. The response to IVIG treatment is better locally in this small study and East Indians seem particularly susceptible to the illness but a community based study was not carried out and only data from two hospitals were reviewed. A larger number of patients need to be studied over a longer period of time to determine if any greater attention needs to be paid to identifying antecedent infections.

REFERENCES

Appendix 1: Diagnostic criteria for Guillain-Barré syndrome (Asbury and Cornblath\textsuperscript{(1)})

I Features required for diagnosis
A Progressive motor weakness of more than one limb
B Areflexia

II Features strongly supportive of diagnosis
A1 Progression within four weeks
2 Relative symmetry
3 Mild sensory symptoms and signs
4 Cranial nerve involvement
5 Recovery within four week of progression stopping
6 Autonomic dysfunction
7 Absence of fever at onset
B1 Raised CSF protein
2 CSF mononuclear leucocyte count less than 10/mm\textsuperscript{2}

C Electrodiagnostic features strongly supportive of the diagnosis (nerve conduction slowing or block)

III Features casting doubt on the diagnosis
1 Pronounced asymmetry of weakness
2 Persistent bladder or bowel dysfunction
3 Bladder or bowel dysfunction at onset
4 More than 50 mononuclear leucocytes/mm\textsuperscript{2}
5 Presence of polymorphonuclear leucocytes in CSF
6 Sharp sensory level

IV Features that rule out the diagnosis
1 Current history of hexacarbon use
2 Abnormal porphrin metabolism
3 Recent diphtheritic infection
4 Features clinically consistent with lead neuropathy
5 Purely sensory syndrome
6 Definite diagnosis of poliomyelitis, botulism, hysterical paralysis, or toxic neuropathy.

Appendix 2: Functional score

0 – Healthy 1 – Minor symptoms and signs 2 – Able to walk more than 10 m without assistance 3 – Able to walk more than 10 m with walker or support 4 – Bedridden or chair-bound. Unable to walk 10 m with walker or support 5 – Requiring assisted ventilation for at least part of the day 6 – Dead