parameters in the TA or BB muscle, although MUPs continued to be significantly abnormal at both time-points (Tables 2 and 3).

Z-scores were calculated for the various multi-MUP parameters (refer to Table S1 in Supplementary Material, available online). BB MUP duration in 91% (29 of 32) of the DMD subjects was >2 standard deviations below that of controls (Z-scores < -2). Similarly, 100% (31 of 31) of DMD subjects had a TA MUP duration of >2 standard deviations below that of controls. Area-to-amplitude ratio was 2 standard deviations below controls for BB and TA MUPs, TA MUP area was also >2 standard deviations below that of controls, but the remaining MUP parameters showed no significant difference.

**DISCUSSION**

In this study QEMG multi-MUP analysis confirmed myopathic changes in all DMD boys. The recorded BB and TA MUPs had significantly reduced amplitude, duration, area, area-to-amplitude ratio, and rise time compared with age-matched healthy controls. Szmidi-Salkowska et al. found subjects with DMD had decreased MUP amplitude in the BB muscle. However, they also noted more polyphasic potentials and increased MUP duration, whereas our study showed no change in polyphasia and demonstrated shorter MUP duration in DMD. The 6-month follow-up QEMG studies did not change significantly, except for the increase in the mean MUP area and amplitude of the TA muscle. However, the area-to-amplitude ratio in the TA muscle remained unchanged on follow-up exam. Possible explanations include the compensatory hypertrophy of the remaining muscle fibers in an otherwise dystrophic muscle condition and, less likely, an EMG needle sampling error.

The lack of change of QEMG parameters in DMD boys was likely due to slow progression of the disease. Given the significantly reduced mean duration of the MUPs at the first EMG examination, the lack of significant change in MUP duration on follow-up study was not unexpected. In addition, functional scores did not change significantly over the 6-month study period. In conclusion, our study suggests that QEMG of the BB and TA muscles is an early and sensitive measure of disease activity in DMD boys. However, it does not reflect disease progression over a short (6-month) interval. Long-term follow-up studies will be helpful in efforts to understand the role of QEMG as a biomarker for disease progression in DMD.

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**AGE MAY CONTRIBUTE TO THE INCREASED FREQUENCY OF AXONAL GUILLAIN-BARRÉ SYNDROME**

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**ABSTRACT:** Introduction: The frequency of axonal Guillain-Barré syndrome (GBS) varies among countries. Previous studies supporting the high frequency of axonal GBS in South America have been carried out with pediatric populations. We seek to determine the frequency of axonal GBS in both children and adults in South America. Methods: This is a retrospective cohort analysis of patients who were diagnosed with GBS between January 2006 and December 2013 in a neurological center in Buenos Aires, Argentina. Adults and children with a diagnosis of GBS were included and classified by applying Ho and colleagues' criteria for axonal GBS. Results: The study included 105 patients with GBS. Among 58 adults, only 5 individuals were classified as axonal GBS compared with 16 of 47 children. The frequency of axonal GBS was significantly higher in children than in adults (34% vs. 8.6%, P = 0.0001). Discussion: As shown in a cohort of South American patients, age may impact the frequency of axonal GBS.
Guillain-Barré Syndrome (GBS) is a potentially life-threatening immune-mediated polyradiculo-neuropathy. It is the most common cause of acute flaccid paralysis worldwide, with a reported incidence of 0.6–4.0 per 100,000 persons per year. GBS was regarded as a demyelinating neuropathy from the time of its original description until an axonal subtype was identified in the early 1990s. Acute inflammatory demyelinating polyneuropathy (AIDP) and the “axonal variants” are now considered the 2 main electrophysiological subtypes of GBS. Acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN) share similar pathological processes and immunological profiles. For this reason, they are grouped together as axonal variants, with AMSAN considered a more severe form of axonal GBS. Because demyelinating and axonal GBS variants have different pathogeneses, clinical features, and patterns of recovery, it has been proposed that they may also have differential responses to treatment.

Since its original description in children and young adults, the variable frequency of axonal GBS has been largely associated with geographic location and levels of sanitation. It is low in North America and Europe (3%–17%), intermediate in Israel (22%) and Japan (38%), and high in Bangladesh (67%) and China (75%). South America and Mexico have an intermediate frequency, accounting for 30%–38% of all cases of GBS. It is remarkable that only 2 pediatric series support this statement and that no data are available on axonal GBS frequency in adult South American populations. Furthermore, previous studies about epidemiology of GBS subtypes did not explore a possible association between axonal GBS frequency and age. This study seeks to determine the frequency of axonal GBS in South American children and adults with similar levels of sanitation.

**MATERIALS AND METHODS**

We conducted a retrospective cohort analysis of patients who had been diagnosed with GBS between January 2006 and December 2013 in our hospital located in Buenos Aires, Argentina. Age, gender, living area, clinical features, diagnostic studies, and treatment were assessed in all cases. Electrodiagnostic studies were reviewed by applying Ho and colleagues’ criteria for axonal GBS (Supporting Information Table 1). The frequency of axonal GBS in adults (>18 years old) was compared with that in children by using a Fisher’s exact test. P < 0.05 was considered significant. Statistical analysis was conducted in Stata V12.1 (Stata-Corp, College Station, TX). The study was approved by the local ethics committee.

**RESULTS.** The overall frequency of axonal variants among 105 patients who had been diagnosed with GBS was 20% (n = 21). Fifty-eight patients (55%) were adults (57% males, median age 49 years, range 20–83 years), and the remaining 47 (45%) were children (53% males, median age 10 years, range 3–17 years). All patients lived in urban or suburban areas with similar levels of sanitation. The median number of days from disease onset (sensory or motor symptoms) to admission was 8, with a range of 3–23 days. Electrodiagnostic studies were performed within the first week of admission in all cases. According to Ho and colleagues’ criteria, only 5 adults (8.6%) had axonal GBS (AMAN), whereas most were classified as AIDP. No patients remained unclassified. On the other hand, 16 children (34%) presented with AIDP and 10% (n = 5) were diagnosed with Miller-Fisher. Two patients (4%) remained unclassified. Axonal variants were significantly more frequent in children than in adults (P = 0.0001).

**DISCUSSION**

In our cohort, we found an overall frequency of axonal GBS of 20%. In children, the frequency was 34%, similar to previous studies in our country and Mexico. However, in adults, the frequency of axonal variants was significantly lower (8.6%) and similar to what has been found in some European countries and North America. Adults and children in our population had similar levels of sanitation. Previous studies indicated that the geographical location, closely related with the level of sanitation, was the main determinant of the probability a priori that a patient would have an axonal GBS variant. Evidence in prior studies on age distribution and its relationship with axonal GBS is scarce. It is noteworthy that those studies that included only adults had the lowest frequency of axonal GBS (summarized in Supporting Information Table 2). On the other hand, studies of only pediatric populations and those in which there was a preponderance of...
children and younger adults showed the highest frequencies of axonal variants. Nonetheless, the role of age has not been specifically addressed until now. Our data suggest that, beside the level of sanitation, age could be a major determinant of the probability of having an axonal versus a demyelinating GBS variant.

*Campylobacter jejuni* infections play a central role in the pathogenesis of axonal GBS. It is well known that the immune response against this bacterium induces the production of antibodies that cross-react with gangliosides present in peripheral nerves, causing direct damage to the axon in axonal GBS variants.3 Infections caused by *C. jejuni*, mostly diarrheas, are also the most important trigger for AMAN and AMSAN, with positive serology in 27%–65% of these patients.3 Thus, we hypothesize that the epidemiology of *C. jejuni* infections could explain, at least in part, the relationship between age and frequency of axonal GBS in our cohort.

*C. jejuni* is a leading cause of diarrhea in children in developing countries, with an estimated incidence of ~30,000/100,000 population. In developed countries, its incidence is markedly lower (~300/100,000 population), possibly related to improved levels of sanitation.9 However, in both cases, it is significantly higher than what is found in adults (~90/100,000).10 These data clearly show that, regardless of the level of sanitation, *C. jejuni* affects primarily children. Also, because *C. jejuni* infections can be asymptomatic, the exposure to it during childhood could be a protective factor against this infection later in life.11

The present study has some limitations. Because some of our patients were referred from other institutions, a referral bias may have been introduced. However, this bias should affect both the adult and pediatric populations equally. In addition, results from a prior study7 in our country confirm the frequency of axonal variants found in our pediatric population. Unfortunately, we were not able to find data on the prevalence of axonal variants of GBS in adult South American populations. The lack of serology against *C. jejuni* is another limitation. This is not routinely performed in our center because it is unlikely to change the management of individual patients.

In conclusion, in our cohort, age seems to have had an impact on the frequency of axonal GBS. We hypothesize that the pathogenic relationship between *C. jejuni* infections and axonal GBS and the epidemiology of *C. jejuni* infections could explain our findings. This should be taken into account and addressed in future epidemiological studies. Our data also show that adult-onset axonal GBS in a South American cohort had a similar frequency compared with those reported in North America and some European countries.

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Ethical Publication Statement: We confirm that we have read the Journal’s position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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RENAL AND HEMATOLOGIC SIDE EFFECTS OF LONG-TERM INTRAVENOUS IMMUNOGLOBULIN THERAPY IN PATIENTS WITH NEUROLOGIC DISORDERS

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ABSTRACT: Introduction: For patients receiving intravenous immunoglobulin (IVIg), renal and hemolytic side effects are well recognized. However, there are very few data on the effects of chronic IVIg therapy. Methods: We retrospectively analyzed laboratory data on 166 patients who received IVIg for 12 months with a dose range of 0.441–2.58 g/kg/month, measuring