ABSTRACT

Sural sparing pattern is believed to be important early neurophysiologic change of Guillain-Barre syndrome (GBS) in adult and it is considered one of the diagnostic criteria for early GBS, but in children, there are limited studies to define its importance. This study aims to find the role of sural sparing pattern in the detection of GBS in children. The study involves 22 children presented with classical features of GBS. Then the diagnosis was confirmed using serial electrodiagnostic testing and CSF examination. Then they undergo full neurological and neurophysiological assessment by nerve conduction study and electromyography. Sural sparing is defined as decrease in the ratio of sensory amplitude between ulnar and sural nerves compared to age and sex matched control. The study found that sural sparing pattern is a sensitive (83%) and specific (92%) finding in GBS. We conclude that sural sparing pattern is useful marker for diagnosis of GBS in children.

Keywords: Guillain-Barre syndrome, flaccid paralysis, sural sparing pattern.

INTRODUCTION

Guillain-Barré syndrome (GBS) is defined as a group of clinical syndromes with acute onset of peripheral neuropathy – axonal or demyelinating – secondary to an immune-mediated process. It is commonly presented as slowly progressive weakness of spinal and less commonly bulbar nerves, with occasional fatal involvement of respiratory muscles (1) and is considered the most common cause of post-infectious paralysis worldwide. The prevalence of the disease is variable between different countries and range from 0.6 to 4 cases per 100,000 (2).
There are three subtypes of GBS depending on the target of pathological process within the nerve. If the pathological process target the myelin sheath it will results in acute inflammatory demyelinating polyradiculoneuropathy (AIDP) which is the commonest subtype. But if the pathological process targets the axon of nerve, it will result in other subtypes like acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN). Other subtype include Miller Fisher syndrome (which patient present with ataxia, areflexia and ophthalmoplegia with minimal or without weakness) (3). These subtypes has different prognosis and outcomes.

Clinical features of pediatric GBS include slowly progressive symmetrical weakness of the limbs which follow ascending pattern in addition to paresthesia, and pain. Autonomic dysfunctions might occur including fluctuating blood pressure, arrhythmia, abnormal sweating, pupillary abnormalities. In addition, one of the serious short-term complication is respiratory failure requiring mechanical ventilation (4).

Early diagnosis of GBS is vital to allow early institution of treatment and prevent disease progression and complications. One of the important tests that can help in the diagnosis of the disease is electrodiagnostic examination (5). However, the electrodiagnostic findings in the early stages of the disease is nonspecific and can’t prove the neuropathic nature of the disease. It is found that electrodiagnostic testing performed within 4 days of onset of symptoms showed nonspecific features of demyelination (like prolonged distal motor latency, abnormal late responses...) in 15 (83%) but no one had shown conduction block or temporal dispersion which are features needed for supporting diagnosis of acute immune mediated demyelination (6).

The goal of electrodiagnostic examination in GBS is to be sure that peripheral nerve dysfunction is the cause of acute weakness which is suspected on clinical ground, to elucidate findings of new onset acquired demyelination like increase temporal dispersion, prolonged distal motor and sensory latencies and F-wave latencies, conduction block, non-uniform slowing of conduction velocities (7). Also electrodiagnostic study help to exclude other causes of weakness like myopathy, motor neuron disease, chronic neuropathy and others. In addition to that, electrodiagnostic characteristics help to predict the prognosis of the disease (8).

Although sensory nerve conduction study is routinely performed with identifiable changes, they are rarely included in the electrodiagnostic criteria (9). One of the most important finding is the “sural-sparing” pattern; which mean, the finding of a normal or relatively preserved sural sensory potential in the presence of abnormal upper limbs sensory nerve action potential (e.g., ulnar, radial or median sensory responses). The importance of sural sparing come from the fact that, it is common finding (seen in approximately 50-70% of patients suspected to have GBS examined early within 7 days of onset of disease) and is very unusual for neuropathies other than GBS (10). So sural sparing pattern is considered to be highly specific marker of GBS (9).

The diagnostic utility of this abnormality has been demonstrated in various studies (11-14). Our recent study aims to define sural sparing pattern as the ratio of amplitudes of ulnar sensory response to sural sensory response and to find the sensitivity and specificity in diagnosing GBS in pediatric patients.
MATERIALS AND METHODS

This is a case control study that involves 22 children studied at the period between 2012 and 2019. They aged between 2-12 years and gender distribution of 9 females and 13 males. Patients presented with clinical features suggestive of GBS like ascending progressive weakness and paresthesia of affecting upper & lower limbs in symmetrical pattern with areflexia or hyporeflexia on clinical examination. The duration of symptoms ranges from 5-14 days. The diagnosis of GBS was confirmed depending on serial nerve conduction studies done after 5-7 days which show progressive demyelination or axonal degeneration compared to old study or examination of cerebrospinal fluid which show increase albumin and sugar with low cellular count.

Nerve conduction study was done for all participants with Nihon Kohden machine 2010. Supramaximal stimulation and standard surface electrodes was used. Latency, amplitude and conduction velocity was tested both motor and sensory fibers of median, ulnar, peroneal, tibial and sural nerves with F-waves and H-reflex testing. Temperature was kept within accepted range throughout the testing. We defined sural sparing as normal sural SNAP amplitude bilaterally compared to two abnormal or absent upper limb SNAP amplitudes either on the same limb or the opposite limb. Total of 22 normal healthy controls also involved in the study. They has the same age and gender characteristics of patient group and also undergo detailed NCS testing.

Statistical analysis was done using SPSS program version 20. Mean and standard deviation of the variables was compared between patients and controls.

RESULTS

Table 1: The demographic data of patients and control

<table>
<thead>
<tr>
<th>Variable</th>
<th>Age (years)</th>
<th>Sex number (male/female)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient</td>
<td>4±6</td>
<td>13/9</td>
</tr>
<tr>
<td>Control</td>
<td>3±9</td>
<td>9/13</td>
</tr>
</tbody>
</table>

The study of motor nerve conduction study show that there is statistically significant differences between patients and control regarding the tested parameters (distal motor latency, amplitude and conduction velocity) as shown in table 2 which demonstrate the findings for both upper & lower limb motor nerves.
The results also outlined statistically significant differences between patients and control group regarding sensory nerves parameters like sensory latency and amplitude. These changes are seen mainly in median and ulnar nerves. While in sural nerve, the results are not significant.

Table 3: The findings of sensory NCS of studied nerves

<table>
<thead>
<tr>
<th>Variable</th>
<th>Patient</th>
<th>control</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Median</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Latency (ms)</td>
<td>2.2±2</td>
<td>1.5±1</td>
<td>0.00</td>
</tr>
<tr>
<td>Amplitude (µv)</td>
<td>16.6±18.8</td>
<td>37±13</td>
<td>0.00</td>
</tr>
<tr>
<td><strong>Ulnar</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Latency (ms)</td>
<td>2.9±1.8</td>
<td>1.8±1</td>
<td>0.008</td>
</tr>
<tr>
<td>Amplitude (µv)</td>
<td>13±15</td>
<td>23±7</td>
<td>0.008</td>
</tr>
<tr>
<td><strong>Sural</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Latency (ms)</td>
<td>2.8±1.7</td>
<td>2±1</td>
<td>0.7</td>
</tr>
<tr>
<td>Amplitude (µv)</td>
<td>12.5±12</td>
<td>11.5±2</td>
<td>0.9</td>
</tr>
</tbody>
</table>

Sural sparing pattern is shown in figure 1. It clearly indicates that sural sparing pattern is seen in 83% of patients and negative in 17%. While it was negative in the control group.
FIGURE 1: The distribution of positive sural sparing pattern in patients with Guillain Barre syndrome.

DISCUSSION

Many previous researches [6,12,15,16] studied the role of sural sparing pattern in the diagnosis of GBS and found that the sensitivity range from 19-67% while the specificity range from 90-100%. Most of these studies are conducted on adult patients, so limited information is present regarding the validity of this test in child age group.

The current study compares between the amplitude of median and/or ulnar nerves sensory responses with those obtained from the sural nerve. Any reduction in amplitude or no response in the upper limb nerves compared to normal sural response is considered as abnormal and positive test. Current study found that the test was positive in 83% of the patients which is higher than the results obtained in adults (range from 19-67%). This variation between adult and children might be related to relatively rapid progression of the disease in children when compared to adults.

The affection of sensory fibers of median and ulnar nerves before those of sural nerve is apparently in contrast with the fact that GBS is an ascending paralytic disease and early manifestation of it start in the lower limbs, but the assumed explanation of the preservation sural nerve in GBS is that the nerve recorded at some distance proximal to its terminal end (behind the lateral malleolus), while median and ulnar sensory nerves recorded at its most terminal end (fingers) and it is well known that GBS affects the most distal segments of the nerves early in the course of the disease [12].

On the other hand, this phenomenon can be demonstrated also as GBS has a predilection to affect entrapment sites and points where the blood nerve barrier is susceptible (for example the median nerve at the wrist and the ulnar nerve at the elbow). Many authors [17] demonstrated diffuse inflammatory demyelination with or without axonal degeneration involves mainly the spinal nerves which are obviously the most proximal portions of the nerves, and so far they are severely affected. This supports the assumption that
disruption of blood nerve barrier (either at nerve roots or at entrapment sites) as a more reasonable explanation for the sural-sparing in GBS(18).

In addition to its role in early diagnosis of GBS, sural sparing pattern can help in (16) the differentiation of GBS from its clinical mimics like chronic inflammatory demyelinating polyneuropathy (CIDP) and diabetic neuropathy. A study done by Bromberg and his study group found that this abnormality occurred in (39%) of GBS patients compared with (28%) CIDP patients and (14%) of diabetic neuropathy sufferers (13).

Other study found that 2 out of our 5 patients with GBS above 70 years displayed the pattern and so they concluded that sural-sparing is useful in older patients too; (19).

The development of sural-sparing has largely been referred to the demyelinating insult of GBS and so it is well documented finding only in the demyelinating form of the disease (AIDP) [11-14].

CONCLUSION

Sural sparing pattern is useful marker for the early diagnosis of GBS in children especially in demyelinating type of disease.

ETHICAL CLEARANCE

The Research Ethical Committee at scientific research by ethical approval of both environmental and health and higher education and scientific research ministries in Iraq

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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REFERENCES


