PERCENTAGE OF PATIENTS WHO SHOW AGREEMENT BETWEEN EMG AND CSF FINDINGS FOR THE DIAGNOSIS OF GBS

FARYAD HUSSAIN, NASIR ABBAS BALOCH, ZAINAB FATIMA, ARIF ZAHEER, PROF AGHA SHABBIR ALI

ABSTRACT
Objective: to determine the percentage of patients who show agreement between EMG and CSF findings for the diagnosis of GBS.
Study Design: Cross sectional survey.
Setting: This study was conducted in the Department of Neurology and Medical ICU, The Children Hospital Lahore, from 01-10-2011 to 31-03-2012.
Methodology: 120 patients fulfilled the inclusion criteria were enrolled in the study. EMG was conducted and conduction velocities of motor and sensory nerves of lower and upper limbs and amplitude of compound motor action potential (CMAP’s) were checked. After ten days of start of disease lumbar puncture was done with aseptic measures by the researcher and was sent to the central laboratory of the hospital for analysis of CSF.
Results: The mean age of the patients was 5.6±2.8 years. There were 72 (60.0%) male patients and 48 (40.0%) female patients. 116 (96.7%) patients had positive EMG findings and 4 (3.3%) patients had negative EMG findings. 66 (55%) patients had positive CSF findings and 54 (45%) patients had negative CSF findings. Thus at least 64 (53.3%) patients had an agreement between CSF and EMG findings.
Conclusion: It is concluded from this study that there is strong agreement between EMG findings and CSF findings for the diagnosis of Guillain Barre Syndrome in children.

Keywords: Guillain Barre Syndrome, EMG findings, CSF findings, agreement

INTRODUCTION
Guillain Barre Syndrome (GBS) is a post infectious polynuropathy involving mainly motor nerves but sometimes also involves the sensory and autonomic nerves. This syndrome affects people of all ages and has been reported throughout the world. Most of the GBS patients have a demyelinating neuropathy but primarily axonal degeneration is documented in some cases. Paralysis usually follow a nonspecific viral infection, 1-3 weeks prior to the onset of weakness. The original infection may occur in gastrointestinal tract (GIT), (especially by campylobacter jejuni, helicobacter pylori) or in respiratory tract (Mycoplasma pneumonae). Vaccines have also been linked to GBS.

The onset is gradual and progresses over days or weeks. Weakness begins in the lower extremities and may involve the trunk, upper limb and finally the bulbar muscles. Initially weakness can cause inability to walk and later to flaccid tetraplegia. Tendon reflexes are lost usually early in the course. Dysphagia and facial weakness are impending sings of respiratory failure. Recovery usually begins 2-4 weeks after the progressive phase.

CSF studies are helpful for the diagnosis. CSF proteins are elevated to more than twice the upper normal limits. Glucose level is normal and there is no pleocytosis. Fewer than 10 WBCs per cubic millimeter are found. There is relatively increased CSF protein (46-300mg/dl) and low cellular count (<10/cmm) in about 80% of patients with GBS. Elevated Cerebrospinal fluid protein is seen in most of the patients particularly after first week of illness due to breakdown of blood nerve barrier within the subarachnoid space.

Motor nerve conduction velocities are greatly reduced and sensory nerve conduction time is often slow during second week of illness. Electromyography (EMG) shows evidence of acute denervation of muscles. EMG is comparable with CSF for diagnosis of GBS But EMG is costly and is not available in many places. On the other hand CSF is a cheaper and readily available modality so it can be used as alternating method where EMG studies can not be done. EMG shows positive findings in 95% of the patients while CSF abnormalities(cytotoalbuminal dissociation) is noted in 52% of the patients. No local studies are available in
the literature so I want to explore the use of CSF examination as an alternative to EMG studies for the diagnosis of GBS in our population.

METHODOLOGY

The calculated sample size is 120 cases with 95% confidence level, 9% margin of error and taking expected percentage of patients i.e. 52% between EMG & CSF findings for diagnosis of GBS. Patients included in study were between 1 to 10 years of age, both male and females and all suspected cases of GBS as per operational definition. Patients who were on artificial ventilation or having signs of meningeal irritation (Neck stiffness, Kernig’s sign, Brudzinski sign) were excluded from the study.

120 patients presenting to the Neurology Department and Medical ICU, The Children Hospital Lahore fulfilling the inclusion criteria were enrolled in the study, after taking an informed consent and biodata. Demographic profile including age, gender and address were recorded. EMG was conducted by a senior Neurologist in The Children’s Hospital in which conduction velocities of motor and sensory nerves of lower and upper limbs and amplitude of compound motor action potential (CAMP’s) were checked using a specific machine by electrical stimulation. After ten days of start of disease lumbar puncture was done with aseptic measures by the researcher and was sent to the central laboratory of the hospital for analysis of positive CSF. All data was entered on a pre-designed Proforma.

Both EMG & CSF findings were interpreted as positive or negative (as per operational definition) for determination of agreement for the diagnosis of GBS. The acquired data was entered and analyzed through SPSS Version 10.0. Variables studied included age and gender. Mean and Standard Deviation was calculated for quantitative variables like age. Frequencies and Percentages were calculated for qualitative variables like gender. Agreement between EMG findings and CSF findings were calculated as frequency and percentage. Kappa statistics was used to determine the strength of agreement between EMG and CSF findings for the diagnosis of GBS.

RESULTS

The mean age of the patients was 5.6±2.8 years. There were 20 (16.7%) patients in the age range of 1.0 to 2.0 years, 26 (21.7%) patients in the age range of 2.1-4.0 years, 26 (21.7%) patients in the age range of 4.1-6.0 years, 21 (17.5%) patients in the age range of 6.1-8.0 years, 27 (22.5%) patients in the age range of 8.1-10.0 years (Table 1). In the distribution of patients by sex, there were 72 (60.0%) male patients and 48 (40.0%) female patients (Table 2). In the distribution of patients by EMG finding 116 (96.7%) patients had positive findings and 4 (3.3%) patients had negative findings (Table 3). In the distribution of patients by CSF finding 66 (55%) patients had positive findings and 54 (45%) patients had negative findings (Table 4). In the distribution of patients with regard to confirmation of agreement, 64 (53.3%) patients had been confirmed as having agreement and 56 (46.7%) patients did not have confirmation of agreement (Table 5). In the distribution of patients by agreement between EMG finding and CSF finding, 116 (96.7%) EMG positive patients and 66 (55%) patients were positive with CSF finding with significant p value of <0.001 (Table 6).

Table 1: Distribution of patients by age (n=120)

<table>
<thead>
<tr>
<th>Age (Years)</th>
<th>No. of patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.0-2.0</td>
<td>20</td>
<td>16.7</td>
</tr>
<tr>
<td>2.1-4.0</td>
<td>26</td>
<td>21.7</td>
</tr>
<tr>
<td>4.1-6.0</td>
<td>26</td>
<td>21.7</td>
</tr>
<tr>
<td>6.1-8.0</td>
<td>21</td>
<td>17.5</td>
</tr>
<tr>
<td>8.1-10.0</td>
<td>27</td>
<td>22.5</td>
</tr>
<tr>
<td>Mean±SD</td>
<td>5.6±2.8</td>
<td></td>
</tr>
</tbody>
</table>

Key:

n Number of patients
D Standard deviation

Table 2: Distribution of patients by sex (n=120)

<table>
<thead>
<tr>
<th>Sex</th>
<th>No. of patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>72</td>
<td>60.0</td>
</tr>
<tr>
<td>Female</td>
<td>48</td>
<td>40.0</td>
</tr>
<tr>
<td>Total</td>
<td>120</td>
<td>100.0</td>
</tr>
</tbody>
</table>

Key: n Number of patients

Table 3: Distribution of patients by EMG findings (n=120)

<table>
<thead>
<tr>
<th>EMG finding</th>
<th>No. of patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive</td>
<td>116</td>
<td>96.7</td>
</tr>
<tr>
<td>Negative</td>
<td>4</td>
<td>3.3</td>
</tr>
<tr>
<td>Total</td>
<td>120</td>
<td>100.0</td>
</tr>
</tbody>
</table>

Key: n Number of patients

Table 4: Distribution of patients by CSF findings (n=120)

<table>
<thead>
<tr>
<th>CSF finding</th>
<th>No. of patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive</td>
<td>66</td>
<td>55.0</td>
</tr>
<tr>
<td>Negative</td>
<td>54</td>
<td>45.0</td>
</tr>
<tr>
<td>Total</td>
<td>120</td>
<td>100.0</td>
</tr>
</tbody>
</table>

Key: n Number of patients
CSF Cerebrospinal fluid

Table 5: Distribution Of Patients By Final Assessment Of Agreement (N=120)

<table>
<thead>
<tr>
<th>Final assessment of agreement</th>
<th>No. of patients</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>64</td>
<td>53.3</td>
</tr>
<tr>
<td>No</td>
<td>56</td>
<td>46.7</td>
</tr>
<tr>
<td>Total</td>
<td>120</td>
<td>100.0</td>
</tr>
</tbody>
</table>

Key: n Number of patients

Table 6: Agreement between EMG finding and CSF finding (n=120)

<table>
<thead>
<tr>
<th>Finding</th>
<th>EMG finding</th>
<th>CSF finding</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. of patients</td>
<td>No. of patients</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Percent age</td>
<td>Percent age</td>
<td></td>
</tr>
<tr>
<td>Positve</td>
<td>116</td>
<td>66</td>
<td>0.001</td>
</tr>
<tr>
<td></td>
<td>96.7</td>
<td>55.0</td>
<td></td>
</tr>
<tr>
<td>Negative</td>
<td>4</td>
<td>54</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3.3</td>
<td>45.0</td>
<td></td>
</tr>
</tbody>
</table>

Key: n Number of patients

DISCUSSION

Although the occurrence of GBS in children is relatively rare, it is the most common cause for the development of acute flaccid paralysis among infants and children. Since the first report of GBS in childhood by Manner-Vinard in 1925, showing an incidence of 0.24–1.26 per 100,000 children under 15 years of age. GBS has had a worldwide distribution which has affected all races and all ages, including the newborn.

In our study the mean age of the patients was 5.6±2.8 years. As compared with the study of Akbayram et al, the mean age of the patients was 5.9±3.8 years, which is comparable with our study.

Gender ratios in individual reports in the literature vary from 1.5 to 2.7 males for one female. The gender ratio in our series was 1.3 in favour of males. The occurrence of GBS in children increases with age, and it is quite rare in children younger than 2 years of age.

In our study there were 60% male and 40% female patients. As compared with the study of Akbayram et al, there were 55.5% male and 44.5% female patients, which is comparable with our study.

CSF is characteristically acellular. Protein levels may be normal during the first week of the illness, but the majority will have an increase in protein if measured 2 or 3 weeks later. Elevated CSF protein concentration in GBS has been mainly associated with increased permeability of the blood–CSF barrier.

CSF studies are helpful for the diagnosis. CSF proteins are elevated to more than twice the upper normal limits. Glucose level is normal and there is no pleocytosis. Fewer than 10 WBCs per cubic millimeter are found. There is relatively increased CSF protein (46-300mg/dl) and low cellular count (<10/cmm) in about 80% of patients with GBS. Elevated Cerebrospinal fluid protein is seen in most of the patients particularly after first week of illness due to breakdown of blood nerve barrier within the subarachnoid space.

Motor nerve conduction velocities are greatly reduced and sensory nerve conduction time is often slow during second week of illness. Electromyography (EMG) shows evidence of acute denervation of muscles. EMG is comparable with CSF for diagnosis of GBS But EMG is costly and is not available in many places. On the other hand CSF is cheaper and readily available so it can be used as alternating method where EMG studies are not available. EMG shows positive findings in 95% of the patients while CSF abnormalities (cytoalbuminal dissociation) is noted in 52% of the patients.

In our study there were 96.7% patients had EMG positive finding. As compared with the study of Akbayram et al there were 95% patients positive with EMG finding, which is comparable with our study.

In our study there were 55% patients had CSF positive finding. As compared with the study of Akbayram et al there were 52% patients positive with CSF positive finding, which is comparable with our study.

The onset is gradual and progress over days or weeks. Weakness begins in the lower extremities and may involve the trunk, upper limb and finally the bulbar muscles. Initially weakness can cause inability to walk and later lead to flaccid tetraplegia. Tendon reflexes are lost usually early in the course. Dysphagia and facial weakness are impending signs of respiratory failure.

Optimal management and treatment of GBS is critically important because the stakes are life or death. Although many patients with GBS are desperately ill and paralyzed, their chances of a full recovery are high if they can overcome the acute stages. Thus, an important aspect of treatment is to provide maximum supportive care during the acute stages. A recent large, multicenter, randomized trial made a comparison between plasma exchange, intravenous exchange and combined treatment. Its final analysis revealed that there was no significant difference in efficacy between these three therapeutic regimens.

The only new observation with patients treated with IVIG was acute relapse in 11.9% of the patients.
relapse rate ranging from 1.4 to 46.7 was reported with use of IVIG. 36,57

Studies of GBS that focused on both children and adults together found that respiratory support was required in about 20–30% of the patients.58,59 Case fatality rates requiring mechanical ventilation for respiratory failure were estimated to be 15–30%.52 Childhood GBS in about one-third of all patients needed ventilatory support for respiratory muscle paralysis, and about 10% of the patients died of the disease and its complications.60

GBS in children has a shorter course and is associated with a more complete recovery than GBS in adult patients. Despite modern treatment regimens, about 10–20% of adult GBS patients continued to be disabled.61,62,63 Moreover, older age at onset was significantly associated with a poorer outcome at 1 year.61 In a retrospective study including adult patients in Taiwan, 12.5% of the patients remained at Hughes scale grade 4–6 after 1 year.64 In contrast, although approximately 40% of the children became nonambulant during their illness and 15–20% required ventilatory support, more than 90% recovered fully, with a small minority showing minimal residual impairment, such as weakness of the ankle dorsiflexor 1–4 months after onset, but were able to walk unaided.65,66 After 1 year, only 14.3% of the pediatric GBS patients needed assistance in walking.52 Moreover, about 72% of the children with GBS could walk independently 1 year after onset, more than twice the percentage of adults.57

In our study, in the agreement between EMG findings and CSF findings, 53.3% patients showed agreement between EMG findings and CSF findings in the diagnosis of GBS. As compared with the study of Akbayram et al52 52% patients showed agreement between EMG finding and CSF finding in the diagnosis of GBS.

On the basis of above discussion it is concluded that there is a strong agreement between EMG and CSF findings for the diagnosis of Guillain Barre Syndrome in children.

CONCLUSION

It is concluded from this study that there is a strong agreement between EMG findings and CSF findings for the diagnosis of Guillain Barre Syndrome in children.

REFERENCES


