Electromyography and Nerve Conduction Study in Predicting Functional Outcome in Paediatric Patients with Guillain-Barre` Syndrome

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A prospective study from January-1999 to March-2001 including 33 consecutive children with Guillain-Barre` Syndrome (GBS) admitted in the Children's Hospital Lahore. Electromyography (EMG) and Nerve Conduction Study (NCS) were performed at 21st day post onset of weakness or earlier, as required to confirm the type of lesion-whether segmental demyelination alone or segmental demyelination along with axonal degeneration. Reduction in conduction velocity, conduction block or abnormal temporal dispersion, prolonged distal motor latencies, absent or prolonged F-wave latencies were indicative of segmental demyelination. The presence of profuse fibrillation potentials in muscles were indicative of axonal degeneration. All patients who underwent in-patient rehabilitation management, were followed up for their neurological and functional status on admission and discharge. The outcome results compared between the two groups. The Modified Functional Independence Measure (MFIM) was used as an assessment tool for evaluating functional status. A good neurological recovery, higher functional independence scores and early discharge was noted in children having segmental demyelination alone. Whereas in children with the axonal variety of the disease who had a more complicated course requiring assisted ventilation at times, poor or delayed neurological recovery and prolonged rehabilitation time were noted. These findings suggest that EMG & NCS is a helpful tool in the diagnosis, prognosis and better management of children with acute GBS.
Key words: Guillain-Barre` Syndrome, Electromyography, Nerve Conduction Study

GBS is a clinical syndrome of progressive muscle weakness with absent reflexes, which develops over a period of three to four weeks usually following a viral or other infection. It affects 1–2 per 100,00 people annually[1-3]. There is an over all annual incidence rate of 0.91/100,000 in children < 15 years with almost 2:1 preponderance towards males[1]. Approximately 10% of the patients die and 20% are left with deficits in ambulation or require ventilator assistance a year later [4, 5].

The acquired nature of the disease, the response to immunotherapy and the pathology all suggest that GBS is an immune mediated disease[5]. However the precise mechanism of immunological injury is unclear. Pathologically, GBS is an inflammatory polyradiculo-neuropathy. The histopathological features include multifocal areas of inflammation and demyelination with cellular infiltration of macrophages and lymphocytes. Approximately 40–60% of patients who have GBS have a history of antecedent illness within previous four weeks[5,6]. Often this illness is upper respiratory tract disease or gastroenteritis. Specific agents implicated include Varicella Zoster, Epstein-Barr, HIV Hepatitis B, Coxsackie viruses, Cytomegalovirus[1,6,7], Mycoplasma pneumonae and Campylobacter jejuni[6]. Campylobacter enteritis has been linked to the more severe axonal variety[9].

The major clinical features are progressive muscle weakness and diminished deep tendon reflexes with symmetrical distribution. Although weakness is the hallmark of the disease, pain and sensory symptoms are present in 50–60% of the patients[10]. Other clinical features that are less constant include cranial nerve involvement with facial weakness, difficulty in swallowing and occasionally involvement of cranial nerves that control ocular motility[11,12]. Autonomic disturbances although infrequent are potentially life threatening with tachyarrhythmias, bradyarrhythmias, hypertension and orthostatic hypotension[13].

The diagnosis of GBS is based on history of acute weakness with findings of objective muscle weakness and areflexia. EMG and NCS is the major diagnostic tool in the investigation of the disease. Classic electrophysiologic findings in GBS confirm the presence of segmental demyelination. The duration of illness is usually less than 12 weeks in the majority of adult cases with most expected to have a favourable outcome (generally this is equated to mean ambulation without assistive devices)[1]. In our experience however, several children with axonal involvement in addition to demyelination, on EMG & NCS had poor neurological and functional recovery. This prompted us to determine whether electrodiagnosis can be a useful tool in the prognostication of acute GBS in children.

Material and methods

The study was conducted at the Children's Hospital & the Institute of Child Health, Lahore from Jan-1999 to March 2001. This hospital is a tertiary care referral center with all types of intensive care and diagnostic facilities. It was a descriptive study including 33 consecutive patients of GBS admitted in the Children's Hospital & the Institute of Child Health, Lahore. Patients included in this study fulfilled the

internationally accepted diagnostic criteria for acute GBS. EMG and NCS was performed with Amplaid 14 machine (M/s Amplifon, Italy) preferably at 21st day post onset of weakness as wallerian degeneration is completed in 3 weeks time. Earlier studies (before 21 days) were however performed on several patients for confirmation of diagnosis to avoid delay in the medical management. Median and Ulnar nerves in the upper and Peroneal and Tibial nerves in the lower extremities were selected for motor nerve conduction study. Tibial F wave study was performed on every case. Sensory study was performed on Median or Sural nerves. EMG of First Dorsal Interosseous or Tibialis Anterior muscle was performed. Reduction in conduction velocity, conduction block, or abnormal temporal dispersion, prolonged distal motor latencies, absent or prolonged F-wave latencies were indicative of segmental demyelination. The presence of profuse fibrillation potentials was indicative of axonal involvement. With the help of Electrodiagnosis the patients were grouped into the demyelinating type or the demyelinating and axonal type. (as illustrated by figures I – VII) All patients were followed up on a regular basis for neurological recovery in both groups. They were provided physical therapy and rehabilitation services during hospitalization (see colour illustration 1 & 2). Modified Functional Independence Measure (MFIM) (see table-I) was utilized to assess ambulation and activities of daily living (ADL) in both groups at admission and discharge and results compared. A performa was also used which was filled up on admission and discharge by the principal investigator based on clinical findings and observations.

Table-I: Modified Functional Independence Measure (MFIM)

a. Locomotion							
Walking	7	6	5	4	3	2	1
Wheel Chair Propulsion	7	6	5	4	3	2	1
b. **Mobility:**							
Wheel chair transfer	7	6	5	4	3	2	1
c. **Self Care.**							
Feeding	7	6	5	4	3	2	1
Grooming	7	6	5	4	3	2	1
Bathing	7	6	5	4	3	2	1
Dressing upper body	7	6	5	4	3	2	1
Dressing lower body	7	6	5	4	3	2	1
Perineal care	7	6	5	4	3	2	1

Key

7 Independent complete.
6 Independent modified.
5 Dependent modified with super vision.
4 Dependent modified with min. assistance.
3 Dependent modified with moderate assistance.
2 Dependent modified with max. assistance.
1 Dependent complete.

Results:

Statistical analysis was completed by chi. square test determining p-values. The following results were obtained in the two groups.

Table II: Important clinical features and recovery factors in GBS

Total no. of cases	Segmental Demyelination (n=15)	Seg. Demyel. + Axonal Degen. (n = 18)	Stat. Analysis (chi sq. test) p value
Pain and paresthesias	9(60%)	16(88%)	< 0.05
Dysautonomia	1(6%)	7(38%)	< 0.05
Ventilatory support*	3(20%)	13(72%)	< 0.01
Plasmapharesis	0	5(27%)	< 0.05
Complete independence in ambulation*	5(35%)	0	< 0.01
Complete dependence in ambulation*	0	11(73%)	<0.01
Complete dependence in ADL*	1(7%)	9(60%)	<0.01
Antecedent illness	12 (80%)	14(77%)	NS
Quadriplegia	5 (33%)	13(72%)	NS
Intravenous Ig	3 (13%)	5(27%)	NS
Areflexia	15(100%)	18(100%)	NS
Sensory loss	0	0	NS
Cranial N. Involvement	1(6%)	1(5%)	NS
Pressure sores	0	0	NS
Muscle contracture.	2 (13%)	1(5%)	NS

Table-III: Functional outcome in patients using Modified Functional Independence Measure (MFIM)

Total no. of cases	Segmental Demyelination (n = 14)	Seg. Demyel. Axonal Degen. (n = 15)
Ambulation		
Independent complete*	5	0
Independent modified*	3	0
Dependent modified with supervision	2	1
Dependent modified with minimal assistance	2	1
Dependent modified with moderate–maximum. Assistance.	2	2
Totally dependent*	0	11
Activities of daily living		
Independent complete*	4	1
Independent modified*	3	1
Dependent modified with supervision	3	0
Dependent modified with minimal assistance	1	2
Dependent modified with moderate to maximum assistance	2	2
Dependent complete*	1	9

Segmental demyelination group expired n=1
Segmental demyelination and axonal degeneration group expired n=3

Discussion

Previous pathologic studies of acute motor axonal neuropathy have shown a strong evidence of the presence of a primary axonal Guillain-Barre` Syndrome (GBS)[14]. In one study by Reisin etal[15] severe reduction of the mean amplitude of the compound muscle action potentials (CAMPs) of motor nerves of children with GBS was used to identify a subgroup of patients with axonal damage that produced more severe weakness and delayed recovery. We studied the compound muscle potentials (CMAPs), F-wave latencies of motor nerves and sensory nerve action potentials (SNAPs) but focused primarily on the presence or absence of denervation potentials on electromyography. The presence of profuse fibrillations potentials in the muscles at rest identified the more severe axonal form of acute GBS in our study. Our results have shown, that a substantial number of children (58%) with acute GBS had the axonal form of the disease. These children had a more complicated course with requirement of ventilatory support (pvalue<0.01). A poor functional outcome in ambulation and activities of daily living (pvalue<0.01) as compared to the demylinating variety was also noted in this group. Thus an early diagnosis of the axonal form of acute GBS in children can be helpful in planning more effective medical management and rehabilitation. In conclusion the above study confirms that electromyogrpahy (EMG) and nerve conduction study (NCS) is a useful tool in the diagnosis, prognosis and better management of children with acute Guillain-Barre` Syndrome (GBS)

Illustration 1: Gait training in parallel bar

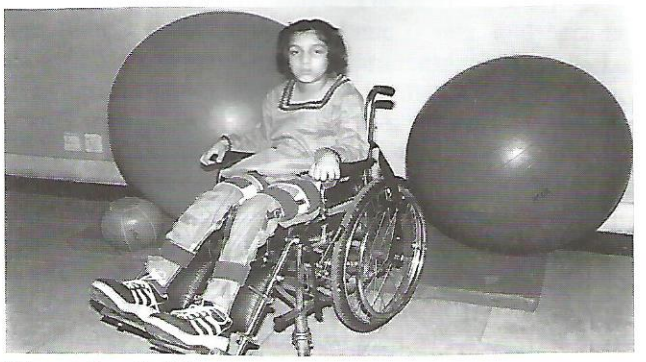

Illustration 2. Wheel chair management and training

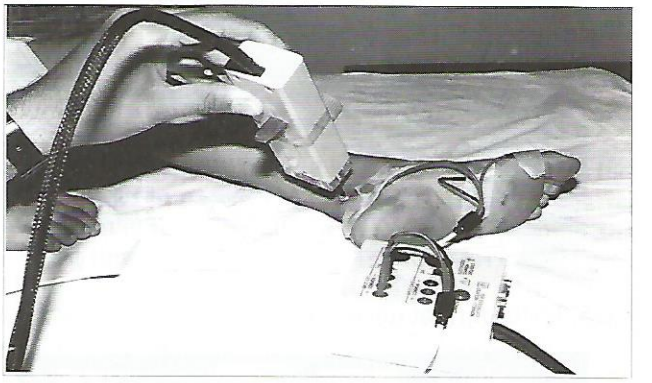

Fig. 1 NCS of left tibial nerve

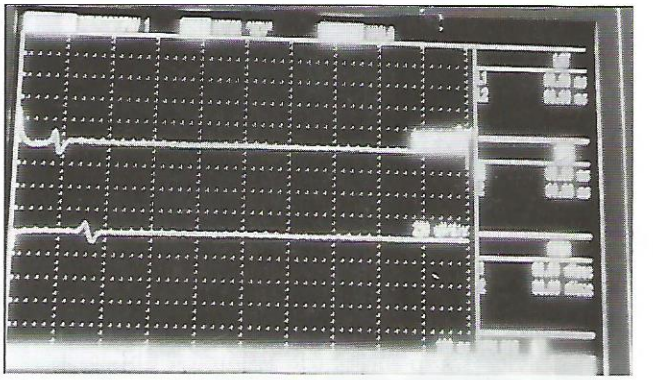

Fig.2 CAMP of tibial nerve

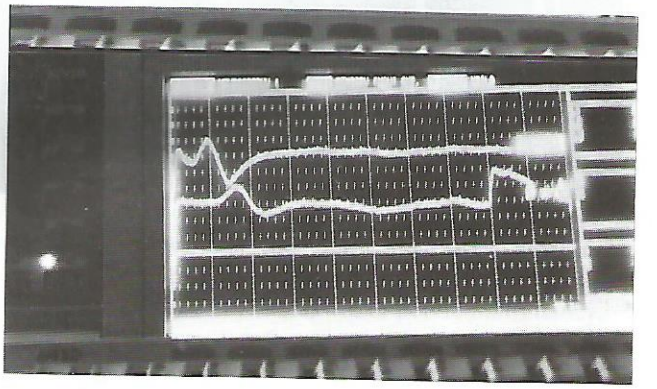

Fig.3 SNAP of median nerve

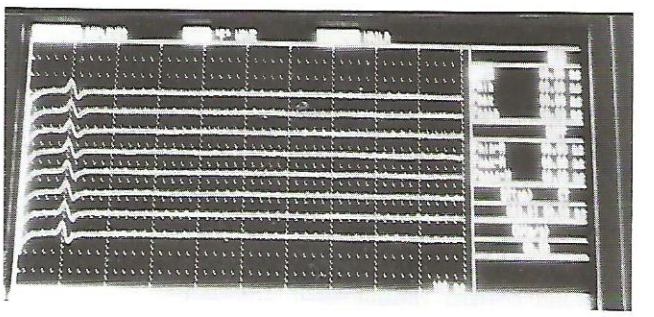

Fig. 4. Tibial F wave study.

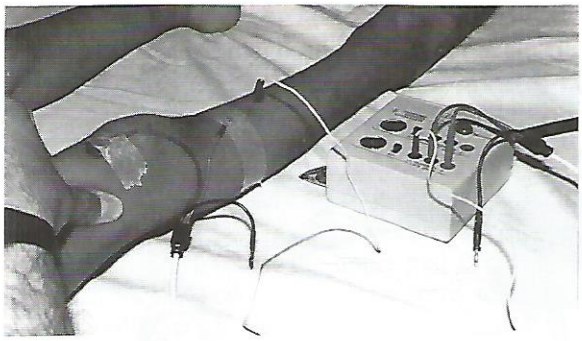

Fig. 5. EMG of right tibialis anterior

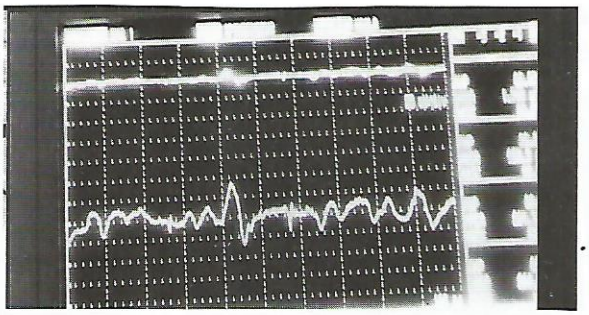

Fig. 6.Fibrillation potentials in muscle at rest.

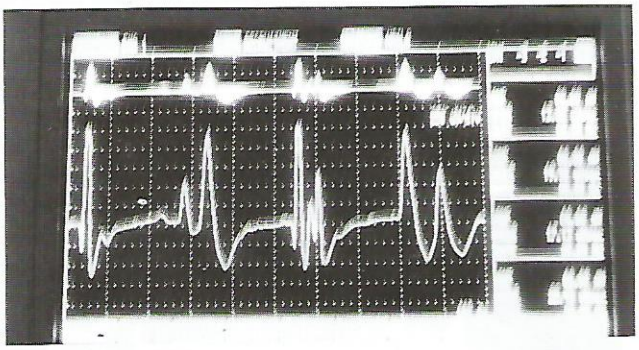

Fig. 7. Motor units action potentials.

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