

An Overview of Guillain-Barre Syndrome with Reference to Cerebrospinal Fluid Analysis and Electrodiagnostic Study at Urban Areas of Madhya Pradesh

Dr. Anand Chandelkar¹, Dr. Arun Chandelkar^{2*}

¹Medical Specialist, Dept. of Gen. Medicine, Govt. District Hospital, Ratlam, India

²Junior Resident, Dept. of Respiratory Medicine, Sri Aurobindo Institute of Medical Sciences, Indore, India

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*Corresponding author

Dr. Arun Chandelkar

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Abstract: Guillain-Barre Syndrome is the commonest cause of acquired demyelinating disorders affecting the peripheral nervous system in any part of the world. It is a spectrum of illness of diverse etiology with a common pathological process. It is a non-seasonal illness affecting persons of all age groups. The severity of Guillain-Barre Syndrome varies from mild weakness to total paralysis and respiratory failure, sometimes leading to death. Predominantly study conducted at Govt. District Hospital, Ratlam & also covered rural population of Ujjain & Indore & its surroundings. It offers medical management to all economic status of people, all religions and all age groups. It is more ideal to conduct a study in this institution. The diagnosis of Guillain-Barre Syndrome is made predominantly by clinical examination and aided by investigations like cerebrospinal fluid analysis and electrodiagnostic studies. 80% of Guillain-Barre Syndrome patients recovered smoothly without going for complications. 30% of Guillain-Barre Syndrome patients developed respiratory muscle weakness of varying severity. 40% of patients showed features of autonomic disturbance of varying severity. 2 patients had features suggestive of ocular muscle involvement and in 1 patient feature of incoordination was present. Prognostic outcome in our study is somewhat poor with increasing age. Prognostic outcome is poor when there is co-existing illness like diabetes mellitus or ischemic heart disease. Cerebrospinal fluid analysis in patients the Guillain-Barre Syndrome who had increased protein correlated with severe demyelination in electrodiagnostic studies and delayed recovery. Prognosis in patients the Guillain-Barre Syndrome linearly varies with severity of electrodiagnostic studies. H-reflex was invariably absent in all those patients included in the study. F-response was absent in 90% of patients in lower limbs and 70% of patients in upper limbs. Reduction in motor nerve conduction velocity was noted in all these patients of varying severity. A feature of conduction block was noted in patients with severe weakness. Recovery was delayed in patients with conduction block than in patients with delayed motor nerve conduction velocity alone.

Keywords: Guillain-Barre Syndrome, Cerebrospinal Fluid Analysis & Electrodiagnostic.

INTRODUCTION

Guillain-Barre Syndrome is the commonest cause of acquired demyelinating disorders affecting the peripheral nervous system in any part of the world. It is a spectrum of illness of diverse etiology with a common pathological process. It is a non-seasonal illness affecting persons of all age groups.

The severity of Guillain-Barre Syndrome varies from mild weakness to total paralysis and respiratory failure, sometimes leading to death.

Predominantly study conducted at Govt. District Hospital, Ratlam & also covered rural

population of Ujjain & Indore & its surroundings. It offers medical management to all economic status of people, all religions and all age groups. It is more ideal to conduct a study in this institution.

The diagnosis of Guillain-Barre Syndrome is made predominantly by clinical examination and aided by investigations like cerebrospinal fluid analysis and electro diagnostic studies.

CLASSIFICATION OF THE GUILLAIN-BARRE SYNDROME SUBTYPES [1-8]

- Acute inflammatory demyelinating polyradiculoneuropathy

- Acute motor axonal neuropathy
- Acute motor sensory axonal neuropathy
- Miller - Fisher Syndrome
- Acute pan-dysautonomia
- Sensory Guillain-Barre Syndrome
- Cervico-brachial-pharyngeal, often with ptosis
- Bilateral facial or abducens weakness with distal paresthesias

Forms of Guillain-Barre Syndrome precipitated by both Campylobacter and Cytomegalovirus show delayed recovery compared to cases unassociated with these two infections [9]. Optic neuritis and pyramidal tract signs are rare manifestations [9].

AXONAL FORMS

- Acute Motor Axonal Neuropathy (AMAN)
- Acute Motor Sensory Axonal Neuropathy (AMSAN)

OBJECTIVES

To assess Guillain-Barre Syndrome with reference to cerebrospinal fluid analysis & electrodiagnostic study.

MATERIALS AND METHODS

Guillain-Barre Syndrome is a monophasic illness; often it is self-limiting. The initial assessment was based on clinical history, detailed neurological examination, routine investigations and special investigations like cerebrospinal fluid analysis and electrodiagnostic studies.

Selection of Patient Criteria

Inclusion Criteria

- Any patient admitted with features suggestive of flaccid progressive weakness affecting all the four limbs were included
- Any patient admitted with progression of weakness of less than 4 weeks duration were included.
- Any patient admitted with reduced or absent deep tendon reflexes were included

Exclusion Criteria

- Any patient admitted with features of hypokalemic periodic paralysis.
- Any patient admitted with features of upper motor neuron signs and symptoms.
- Any patient admitted with severe protopathic sensory symptoms.
- Any patient admitted with history of bite preceding the illness.
- Any patient admitted with history of exposure to toxins like organophosphates.
- Any patient with severe terminal illness.
- Patients admitted with history of suspected food poisoning.

- Patients in whom the weakness progressed for more than 4 weeks Number of cases studied: 50

Duration of study: Jan 2016 to July 2017.

Detailed neurological examination including higher mental functions, cranial nerves, motor system, sensory system and autonomic system was done for all 50 patients.

Autonomic dysfunction was looked for in all these patients. History of dryness of mouth, postural giddiness and defective sweating over the body were specifically asked for.

Blood pressure was routinely taken in lying and sitting posture and if possible in standing posture to bring out orthostatic hypotension. Sympathetic skin response was not done due to technical problem.

Respiratory function tests were done in all patients, everyday during hospitalization, including breath-holding time, single breath count, blowing candle at one arm length, chest expansion, Litten's phenomenon.

Likewise, basic investigations like complete blood count, peripheral smear, blood sugar and urea, serum creatinine and electrolytes, erythrocyte sedimentation rate, daily electrocardiogram, chest x-ray were done for all the 50 patients. Lumbar puncture was done for 42 patients and Cerebro Spinal Fluid was sent for Gram's stain, biochemical and cytological analysis.

Electrophysiological studies were conducted by using the machine of Nerve conduction were done in both upper and lower limbs.

In upper limbs, proximal latency, distal latency, motor nerve conduction velocity, F-response were studied in ulnar, median and radial nerves.

In lower limbs, similarly proximal latency, distal latency, motor nerve conduction velocity, F-response, H-reflex were studied in sciatic, lateral popliteal and posterior tibial nerves. Sensory conduction velocity studied in median nerve, ulnar nerve and sural nerve.

Electromyography was done with surface electrodes in thenar and hypothenar muscles, quadriceps, calf muscles, extensor digitorum.

Insertional activity was recorded Resting activity was recorded. Fibrillation potential, fasciculation potential, positive sharp waves were looked for.

Recruitment and interference pattern were looked for. Compound muscle action potential was recorded.

Magnetic Resonance Imaging was done in 4 patients who presented with altered sensorium, sensory disturbance and urinary retention.

OBSERVATION & DISCUSSION

In the 50 patients studied, 30 were male and 20 were female.

CEREBRO SPINAL FLUID ANALYSIS

Cerebrospinal fluid analysis was done for 42 patients. Cerebrospinal fluid protein was increased in all the 42 patients ranging from 260mgs% to 1.5 gms%. Cerebrospinal fluid cell count was within normal limits, i.e. less than 50 mononuclear cells per cubic millimeter. Cell count was 250 in one patient with HIV infection. CSF analysis could not be done in 8 patients due to various reasons.

Table-1: Cell Counts In Patients

CELL COUNT per cubic mm	<10	10-20	21-30	31-40	41-50	>50
No. of Patients	5	9	19	7	1	1

Table-2: Electrodiagnostic Studies Nerve Conduction Studies Upper Limbs

	No. of Patients	Ulnar	Median	Radial
Proximal latency	37	Prolonged	Prolonged	Prolonged
	5	Normal	Normal	Normal
Distal latency	40	Prolonged	Prolonged	Prolonged
	2	Normal	Normal	Normal
MNCV	36	Delayed	Delayed	Delayed
	6	Normal	Normal	Normal
F-Response	30	Absent	Absent	Absent
	12	Prolonged	Prolonged	Prolonged
SNCV	12	Absent	Absent	-

Table-3: Nerve Conduction Studies Lower Limbs

	No. of Patients	Common Peroneal	Posterior Tibial	Sural
Proximal latency	40	Prolonged	Prolonged	
	10	Normal	Normal	-
Distal latency	45	Prolonged	Prolonged	
	5	Normal	Normal	-
MNCV	43	Delayed	Delayed	
	7	Normal	Normal	-
F-Response	42	Absent	Absent	
	8	Prolonged	Prolonged	-
H-Reflex	50	Absent	Absent	-
SNCV	12	-	-	Absent

The disease progressed in 40 patient's up to 14 days. 35 patients worsened in the first week. 15 patients worsened after the first week.

40 patients had shown some improvement during hospital stay itself. 10 patients did not show any improvement during hospital stay. Among 40 patients in whom improvement had seen, recovery was rapid during the first week in 35 patients.

Cerebrospinal fluid analysis and electrodiagnostic studies were done only once during their hospital stay and follow up study could not be done.

Magnetic resonance imaging was done in 4 patients which showed no evidence for cord compression or myelitis.

Table-4: Types Of Guillain-Barre Syndrome

Types	No. of Patients	Percentage
AIDP	25	50%
AMAN	12	24%
AMSAN	10	20%
MFS	1	2%
SENSORY GBS	2	4%

DISCUSSION

GBS seems to affect all the age groups. However there are studies suggesting some more specific age distribution. Adams, Victor and Ropper [1] have quoted age range from as small as 8 months to as old as 80 years for GBS.

Kaplan et reported two peaks one between 15 to 35 years and the other between 50 to 75 years of age. Jiang-Guoxin *et al.*, [10] in their study in

Sweden have observed two peaks one between 20 to 24 years and the other between 70-75 years. In our study among the 50 patients studied, 52 % (26) of patients were below

40 years, 30% (15) of patients were between 40-60 years and 18% (9) of patients were above the age of 60. The age range of our consecutive patients has been 16 years to 78 years, with attack rates highest in persons less than 50 years of age

Table-5: Comparison of CSF analysis:

Name of the study	Percentage
Winter <i>et al.</i> , [11]	80%
Ropper <i>et al.</i> , [1]	90%
Yang <i>et al.</i> , [15]	84%
Present study	85%

Steroid Use

Most of the studies do not recommend the use of steroids. Yet many centers use high dose oral prednisolone or methylprednisolone. In our hospital, we do not use steroids routinely, some patients included in our study, had been treated with steroids outside. 6% (3) of patients on steroids developed peptic ulceration and gastrointestinal bleeding, with delayed recovery.

2% (1) of patients with diabetes mellitus who were put on steroids developed ketoacidosis and turned out with delayed recovery.

In our study, there was no significant difference in the outcome of patients treated with or without steroids, similar to the word literature. A randomized trial of oral prednisolone therapy by Guillain-Barre Syndrome steroid trial group in 1993, showed no benefit. Pangariyaeta [16] showed no benefit in guillain barre syndrome. A study by Hughes et al 1978, suggested that steroids might increase the subsequent relapse rate.

Electrode Diagnostic Study

All these 50 patients had abnormal electro diagnostic studies like

- Motor Nerve Conduction Velocity was decreased in all the 4 limbs varying from mild to severe degree.

- Conduction block was present in 6 patients.
- H- Reflex could not be elicited in all the patients.
- F- Response was prolonged in 8 patients and absent in 42 patients.
- Compound Muscle Action Potential was decreased in more than 40 patients.

Prognosis in patients with Guillain-Barre Syndrome varied linearly with severity of demyelination or axonal degeneration detected by electrodiagnostic studies. Recovery was delayed in patients with conduction block than in patients with delayed motor nerve conduction velocity alone.

Recovery was earlier and favorable in patients with absent H-reflex and F- response and delayed motor conduction than in patients with conduction block. Adistal CMAP amplitude of less than 20% of the lower limits of normal was associated with poor outcome in the North American Guillain-Barre syndrome study group

Case fatality

In our study, mortality was around 10 percent. Commonest mortality were cause of Respiratory failure and ventricular arrhythmias.

Table-6: Comparison of case fatality

Name of the study	Percentage
Winter <i>et al.</i> , [11]	13%
Singh <i>et al.</i> , [12]	8%
Hughes <i>et al.</i> , [13]	6%
Smith <i>et al.</i> , [14]	4%
Present Study	10%

Poor Prognostic Factors

In a study by Winer *et al.*, [11] age greater than 40 years was found to be a significant prognostic factor. Singh *et al.*, [12] also found that age did not affect outcome. In our study, greater age was found to be a poor prognostic factor.

A poor prognosis has been observed in some studies, in patients having a rapid progression of weakness, a prolonged period of peak paralysis (Plateau) and a delayed onset of recovery, not commencing within 3 weeks from onset of weakness. Winer *et al.*, [11] in a retrospective study of 71 patients, noted that a prolonged plateau time and a failure to improve within 3 weeks were associated with poor prognosis. Our study also showed same results.

Study by Singh *et al.*, [12] shows that presence of bulbar paralysis is also associated with a poor prognosis. Our study also had poor prognosis with bulbar weakness.

The need for assisted ventilation has also been found to be a significant prognostic factor in our study which is in accord with the prognostic studies of both Singh *et al.*, and Winer *et al.*, [11] Our study also showed patients on mechanical ventilator has poor prognosis

Cerebrospinal fluid analysis in patients with Guillain-Barre Syndrome who had increased protein correlated with severe demyelination in electrodiagnostic studies showed delayed recovery.

CONCLUSIONS

- There was male preponderance in our study.
- 80% of Guillain-Barre Syndrome patients recovered smoothly without going for complications.
- 10% of patients developed bulbar weakness of varying severity.
- 30% of patients developed neck muscle weakness of varying severity.
- 30% of Guillain-Barre Syndrome patients developed respiratory muscle weakness of varying severity.
- 10% of patients needed ventilatory support to maintain oxygen saturation.
- 40% of patients showed features of autonomic disturbance of varying severity.

- 2 patients had features suggestive of ocular muscle involvement and in 1 patient feature of incoordination was present.
- Prognostic outcome in our study is somewhat poor with increasing age.
- Prognostic outcome is poor when there is co-existing illness like diabetes mellitus or ischemic heart disease.
- Cerebrospinal fluid analysis in patients the Guillain-Barre Syndrome who had increased protein correlated with severe demyelination in electrodiagnostic studies and delayed recovery.
- Prognosis in patients the Guillain-Barre Syndrome linearly varies with severity of electrodiagnostic studies.
- H-reflex was invariably absent in all those patients included in the study. F-response was absent in 90% of patients in lower limbs and 70% of patients in upper limbs. Reduction in motor nerve conduction velocity was noted in all these patients of varying severity. A feature of conduction block was noted in patients with severe weakness. Recovery was delayed in patients with conduction block than in patients with delayed motor nerve conduction velocity alone.
- In our study, there was no significant difference in the outcome between patients treated with or without steroids.
- Mortality was around 10% in our study.
- The commonest cause of mortality was respiratory failure and fatal ventricular arrhythmias.

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