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# Discovering an Expeditiously Accelerating Case of Motor Neuron Disease Electrophysiologically: A Case Report

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## ABSTRACT

Motor neuron diseases (MNDs) are rare, progressive neurodegenerative disorders affecting upper and lower motor neurons, causing muscle weakness, wasting, and disability. Subtypes include ALS and spinal muscular atrophy. Diagnosis involves clinical evaluation and electrodiagnostic testing. Though incurable, interdisciplinary care improves survival and quality of life. Onset typically occurs at 55–65 years. We report a case of a 25-year-old male who presented with rapidly progressive weakness, muscle wasting, and fasciculations evident in her tongue. Electrophysiological studies revealed evidence of motor neuron disease. This case highlights the aggressive nature of MND and the importance of early diagnosis and palliative care.

***Keywords:*** Motor neuron disease, Nerve conduction study, Electrophysiology

## INTRODUCTION

Motor neuron diseases (MNDs) are a group of rare neurodegenerative disorders showing the signs of both the upper and lower motor neurons affecting the voluntary motor system by progressive muscle weakness, wasting, and fasciculations[1,2,3]. They involve motor neurons in the frontal cortex, anterior horn cells, cranial motor nerve nuclei, and the corticospinal and corticobulbar tracts[1,2,3]. MND can be acquired or hereditary[1]. The disease is often fatal, with a median survival time of 2-5 years from symptom onset. It leads to progressive disability, characterized by limb weakness, breathing difficulties, and problems with speech and swallowing[4]. This group includes amyotrophic lateral sclerosis (ALS), primary lateral sclerosis, hereditary spastic paraplegia, progressive muscular atrophy, spinal muscular atrophy, and pseudobulbar palsy [3]. Globally, the prevalence is around 4.5 per 100,000 individuals, with an annual incidence of 2 per 100,000[4]. According to the 2016 Global Burden of Disease (GBD) estimates, the incidence rate of MNDs across all ages was 0.78 per 100,000 person-years. Furthermore,

the age-standardized prevalence was notably higher in high-income regions such as Europe, Australia, and North America, excluding the Asia-Pacific region[3]. The average age of onset ranges from 55 to 65 years, and the condition is more common in males[4].

The history and examination characteristics of each patient determine the best course of action for diagnosing MND. Although evaluation may include genetic testing, spinal fluid analysis, blood work, targeted imaging of the neurological system, or other modalities, electrodiagnostic (EDX) testing is essential for establishing an early, precise diagnosis[1]. Nerve conduction studies (NCS), needle electromyography (EMG), and repeated nerve stimulation (RNS) are standard clinical techniques for evaluating the motor unit[2]. While no cure exists, advances in interdisciplinary care, including non-invasive ventilation, nutritional support, and symptomatic treatments, have improved quality of life and survival. Medication for the symptoms of spasticity and excessive salivation may be beneficial[5].

## CASE REPORT

A 25-year-old female presented with a history of progressive weakness and muscle wasting in her arms, hands, and legs bilaterally. She reported difficulty with fine motor tasks, such as putting on and off her clothes, and difficulty walking due to weakness in his legs. She also reported difficulty in speaking, and swallowing, and increased fatigue. Fasciculations were evident in her tongue and Nasalis muscle. Facial muscle weakness with positive Bell's Phenomenon was also noted bilaterally.

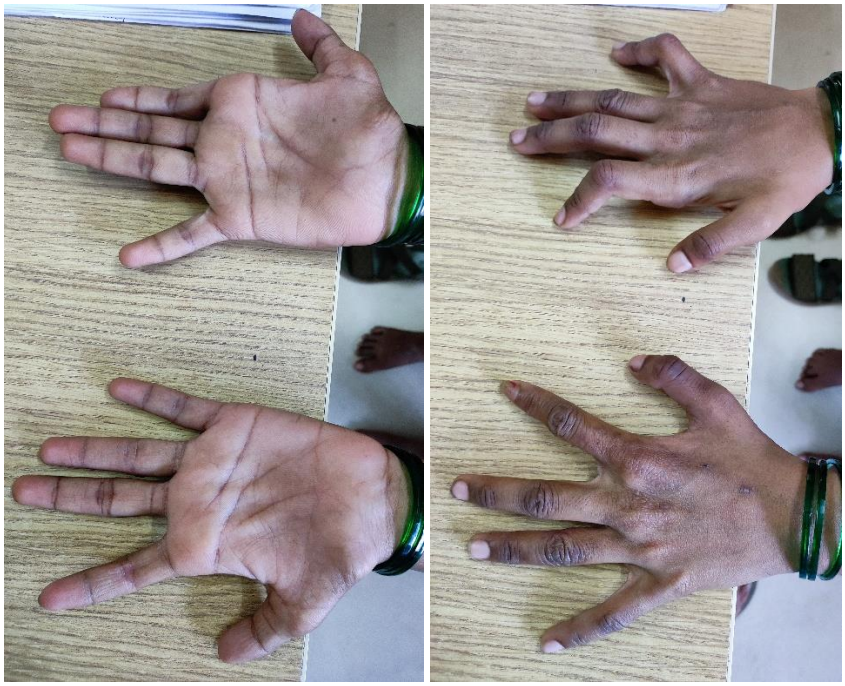


**Fig 1: Split Hand Syndrome: (A) Palmar View, (B)Dorsal View**

Profound atrophy was found over the thenar and hypothenar eminences, palm bilaterally, with clumsiness of the hands, and absent heel strike during gait. A split hand syndrome was quite evident revealing dissociated hand muscle weakness. Both dysphagia and dysarthria with decreased volume of speech were noted. On evaluation of muscle power distal small muscles were more involved than proximal muscles.

### Interpretation of NCV

The motor nerve conduction studies [MNCV] of the Bilateral Median, Ulnar Tibial, and Deep Peroneal, nerves were abnormal. However, the sensory nerve conduction studies of the nerves of the

bilateral Upper limb & lower limbs were found to be within normal limits. The Motor nerve conduction velocities of the bilateral Median, Left Ulnar, & Right Radial nerves were reduced. The onset latency of the Bilateral Median. Right Radial, Left Ulnar & Right Deep Peroneal nerves were prolonged with a marked reduction of the amplitude of compound motor action potential (CMAP) suggestive of both demyelination and axonal degeneration.

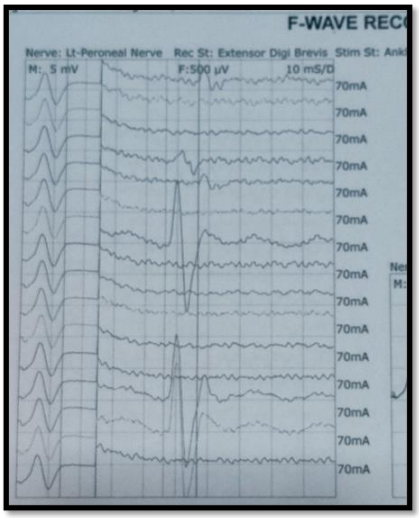



**Fig 2: Increased amplitude of F-waves**

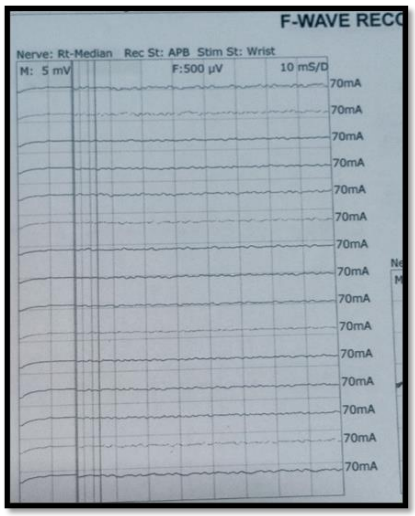
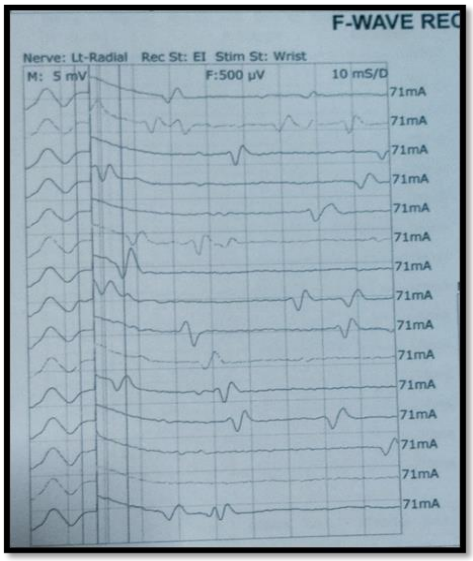



**Fig 3: Absent F waves in Median nerve**



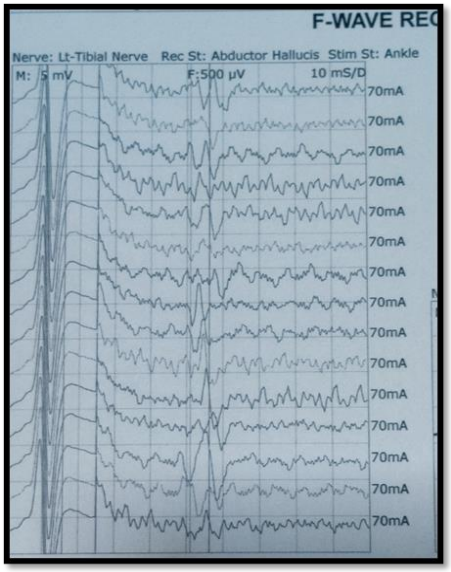




**Fig 4 & 5: Repeater F- waves**

F-wave studies were found to be abnormal. The amplitude of F-waves was found to be increased. F-waves were absent in bilateral Median nerves. Increased chrono dispersion of F waves was found in the Left Radial, Left Peroneal, Right Tibial & Right Ulnar nerves [Fig 2 & 3]. F-repeater waves were evident findings that are primarily related to lower motor neuron loss in ALS [Fig 4& 5].

Bilaterally reduced nerve conduction velocities of the Median, Left Ulnar, Tibial, and Deep peroneal nerves with decreased CMAPs and prolonged onset latencies in motor nerves, abnormal F wave studies were suggestive of Suspected Motor neuron disease [ALS] [Table 1].

| Type of Study | Side/Nerve | Stimulation Site | Recording site | Distance | Latency | Amplitude | Conduction velocity |
|---|---|---|---|---|---|---|---|
| | Left Median | Wrist | APB | 21 cm | 5.83 ms | 28.2 µV | 28.00 m/s |
| | | Elbow | | | 13.33 ms | 0.5 mV | |
| | Right Median | Wrist | APB | 20 cm | 5.31 ms | 0.3 mV | 40.00 m/s |
| | | Elbow | | | 10.31 ms | 0.8 mV | |
| Motor | Left Ulnar | Wrist | ADM | 19 cm | 3.13 ms | 7.9 mV | 52.05 m/s |
| | | Elbow | | | 6.77 ms | 7.1 mV | |
| | Right Ulnar | Wrist | ADM | 22 cm | 3.33 ms | 9.5 mV | 61.97 m/s |
| | | Elbow | | | 6.88 ms | 10.3 mV | |
| | Left Radial | Forearm | Extensor Indicis | 18 cm | 3.33 ms | 3.5 mV | 61.64 m/s |
| | | Above Elbow | | | 6.26 ms | 2.7 mV | |
| | Right Radial | Forearm | Extensor Indicis | 23 cm | 5.42 ms | 1.6 mV | 49.15 m/s |
| | | Above Elbow | | | 10.10 ms | 0.9 mV | |
| | Left Tibial | Ankle | Abductor Hallucis | 34 cm | 6.77 ms | 33.4 mV | 37.95 m/s |
| | | Knee | | | 15.73 ms | 23.6 mV | |
| | Right Tibial | Ankle | Abductor Hallucis | 36 cm | 6.04 ms | 20.4 mV | 36.00 m/s |
| | | Knee | | | 16.04 ms | 12.8 mV | |
| | Left Deep peroneal | Ankle | EDB | 33 cm | 3.44 ms | 6.8 mV | 34.01 m/s |
| | | Knee | | | 12.92 ms | 5.3 mV | |
| | Right Deep peroneal | Ankle | EDB | 32 cm | 4.79 ms | 1.3 mV | 22.92 m/s |
| | | Knee | | | 18.75 ms | 0.7 mV | |
| Sensory | Right Median | Wrist | Dig 2 | 12 cm | 1.63 ms | 53.3 µV | 74.07 m/s |
| | Left Median | Wrist | Dig 2 | 14 cm | 1.63 ms | 171.3 µV | 86.42 m/s |
| | Right Ulnar | Wrist | Dig 5 | 12 cm | 1.96 ms | 252.7 µV | 61.22 m/s |
| | Left Ulnar | Wrist | Dig 5 | 10 cm | 1.83 ms | 163.2 µV | 54.64 m/s |
| | Right Sural | Mid-calf | Ankle | 17 cm | 2.38 ms | 148.6 µV | 71.43 m/s |
| | Left Sural | Mid-calf | Ankle | 19 cm | 2.88 ms | 227.2 µV | 65.97 m/s |
| | Right Superficial Peroneal | Lateral Malleolus | Popliteal Fossa | 11 cm | 1.50 ms | 116.1 µV | 73.33 m/s |
| | Left Superficial Peroneal | Lateral Malleolus | Popliteal Fossa | 15 cm | 3.29 ms | 137.5 µV | 45.59 m/s |

**Table 1: Nerve conduction studies: Bilateral Median, Ulnar, Deep Peroneal, Superficial Peroneal, Sural and Tibial nerve**

## DISCUSSION

This case highlights the aggressive nature of MND, its salient clinical features, and role of electrodiagnosis in confirming the diagnosis.

Amyotrophic lateral sclerosis (ALS) is a progressive and inevitably fatal neurodegenerative disease belonging to a heterogeneous group of disorders known as motor neuron diseases. ALS is the most common adult MND worldwide and is considered the prototypical disorder of this class[6]. Assessing muscle atrophy and tone and testing for abnormal muscle stretch reflexes in all four anatomic regions - bulbar, cervical, thoracic, and lumbar - should necessarily be included in the physical examination[6,7,8]. Though there is variation in clinical presentation, the majority of the patients present with asymmetric limb weakness (80%) or bulbar dysfunction (20%). Bulbar dysfunction can manifest as dysphagia (trouble swallowing) and dysarthria (trouble speaking)[9]. The above said dysfunction was evident in our study patient too. The "split-hand" phenomenon denotes dissociated muscle weakness preferentially affecting muscles in the proximity of the thenar region as compared to the hypothenar muscles of the hand, and has been shown to be a specific electrodiagnostic sign for diagnosing ALS [Fig 1 ][10,11]. However, the frequency of this finding as well as the impact of onset region in relation to split-hand phenomenon remains relatively unknown[11].

Because prognosis and survival can be vastly different among the motor neuron disorders, differentiating among these disorders and making an accurate and timely diagnosis are paramount[6]. While late-stage disease is relatively easy to identify, early disease can be much more difficult to diagnose on clinical examination alone. It is in this situation, early in disease when LMN abnormalities may be hidden from clinical appreciation, that electrodiagnostic evaluation becomes most valuable. Electrodiagnostic testing is considered an extension of the physical examination and should be performed on all patients suspected of having an MND.

Nerve conduction studies (NCS) and needle electromyography (EMG) are collectively termed 'clinical neurophysiology'. They enable the clinician to detect signs that cannot be confirmed by neurological examination alone and can guide diagnosis and treatment [12,13].

The MND electrodiagnostic evaluation includes peripheral NCS examining at least one upper and one lower limb and should begin by evaluating the most severely affected sites.[6,8,14,15] A complete evaluation includes motor, sensory, and F-wave nerve conduction studies. *Motor conduction studies* are an essential part of the electrodiagnostic evaluation of a patient suspected of having an MND. These studies allow the exclusion of treatable neuropathies, such as multifocal motor neuropathy with conduction block, from the differential diagnosis and should include proximal stimulation sites (above the elbow and Erb's point) to rule out conduction block and temporal dispersion, which are not typical findings in ALS [14,15].

Common findings observed from motor NCS in patients with MND include asymmetric side-to-side CMAP differences, normal CMAPs, or CMAPs with decreased amplitude, prolonged distal motor latency, and slowed conduction velocity consistent with axon loss [14,16,15]. These findings were depicted in our study with MND patients.

In addition, distal motor latencies and slowing of conduction velocity worsen as the severity of muscle weakness increases.[18] These findings have been attributed to slowly conducting distal regenerating motor axons and loss of the fastest conducting lower motor neurons[19,20]

**F-Wave Studies:**

Several studies have reported abnormalities in F waves of patients with ALS.[8,19,20]. F-wave latencies and Chrono dispersion, defined as the difference between the maximal and minimal F-wave latencies, were increased in the ALS patients as

compared to the controls in this study by Argyriou and colleagues[19]. Similar findings of F repeater waves and increased amplitudes of F waves were evident in this study.

## CONCLUSION

A definite electrodiagnosis of MND requires demonstration of denervation in four body regions (or three body regions including bulbar, cervical, and lumbosacral) with lack of conduction block and normal sensory potentials along with detailed neurological evaluation.

### *Declaration by Authors*

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