

**Clinico-Epidemiological Study of Motor Neuron Disease with Special Emphasis on
Neurophysiological Diagnostic Aspects**

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Abstract

Motor **N**eurone **D**isease (MND) is a neurodegenerative condition characterised by the progressive weakening of bulbar, thoracic, leg, and abdominal muscles. Both Nerve Conduction Studies (NCS) and Needle Electromyography (EMG) serve as an expansion of the neurological examination and are recommended to be conducted when there is suspicion of a motor neuron disorder. **The present investigation is a** cross-sectional study conducted in Basrah city, southern Iraq. The study involved 18 patients with confirmed MND diagnoses. Their epidemiological, clinical, and electrophysiological characteristics using nerve conduction studies and needle electromyography have been registered. **The** study **aimed at** to **elaborating the** electrophysiological changes among patients with motor neurone disease in Basrah, in addition to some clinical features and epidemiological risk factors.

Keywords: **MND; EMG; NCS;** motor neurone disease; neurophysiology.

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Introduction

Motor **N**euron **D**isease (MND) is a neurodegenerative condition characterised by the progressive weakening of bulbar, thoracic, leg, and abdominal muscles while sensory function remains unaffected. Typically, respiratory failure is associated with a mortality rate of around two to five years. Approximately 85–90% of instances of MND are classified as sporadic, whereas the remaining 10% are attributed to family factors. The global incidence rate of MND was found to be 5 cases per 100,000 individuals. ⁽¹⁾

From a clinical perspective, MND is characterised by the manifestation of muscular atrophy and weakness. The onset of symptoms usually occurs at a specific anatomical location and then spreads, leading to the progression of the illness. Eventually, the patient experiences a loss of voluntary control over skeletal muscles, resulting in a shift from a state of independence to dependency. During the advanced phases, the use of mechanical ventilation and the provision of nutritional assistance may become necessary. The characteristic distribution of symptoms seen in individuals with MND is deemed essential for the accurate identification of this condition. ⁽²⁾ While there is variability in the clinical manifestation, the predominant proportion of individuals exhibit asymmetrical limb weakness (80%) or bulbar dysfunction (20%). Bulbar dysfunction may present as dysphagia or dysarthria. The condition exhibits a gradual dissemination to further anatomical regions, accompanied by concurrent upper motor neuron and lower motor neuron manifestations. ⁽³⁾ Upper motor neuron manifestations include spasticity, heightened reflexes, and a positive Babinski sign. Clinical manifestations of lower motor neuron involvement include several characteristics, such as muscular atrophy, weakness, flaccid paralysis, absent reflexes and fasciculations. ⁽⁴⁾

Electrodiagnostic testing is necessary if the suspicion of MND arises. The assessment of lower motor neuron integrity using electrodiagnostic testing is a critical component in the diagnosis of motor neuron illness, particularly when neuroimaging and laboratory procedures provide

normal results. Nerve conduction studies (NCS) and needle electromyography (EMG) play a crucial role in the diagnostic process of MND by providing valuable help in confirming the presence of the condition and excluding other conditions that may resemble its symptoms. Several illnesses, such as multifocal motor neuropathy with conduction block, chronic inflammatory demyelinating polyradiculoneuropathy, central nervous system tumours, multiple sclerosis, and polyradiculopathy, among others, have the potential to imitate the symptoms of motor neuron disease. ⁽⁵⁾

The use of electrodiagnostic testing is of utmost importance in the diagnostic process of MND due to its ability to identify and assess the presence of lower motor neuron impairment. Both NCS and needle EMG serve as an expansion of the neurological examination and are recommended to be conducted when there is suspicion of a motor neuron disorder. Due to the significant consequences associated with identifying a patient with MND, it is crucial to use NCS and needle EMG to effectively exclude other conditions that may resemble motor neuron disease. Multifocal motor neuropathy with conduction block (MMN-CB) is an illustrative instance of an illness that exhibits significant similarities to MND and necessitates a thorough exclusionary assessment. ⁽³⁾ Similar to MND, it often manifests as gradually developing asymmetrical weakness, but sensory nerves remain unaffected. Typically, the motor nerves of many nerves are impacted. In cases of MMN-CB, it is often seen that there is a notable degree of muscular weakness in comparison to muscle atrophy. Additionally, there is a lack of upper motor neuron manifestations and a greater degree of upper extremity involvement, with a corresponding absence of bulbar findings. ⁽⁶⁾

The El Escorial criteria have been used to standardise the diagnostic criteria for MND. According to the El Escorial criteria, it is necessary to see clinical manifestations of upper motor neuron symptoms, lower motor neuron signs, and a gradual dissemination of signals from one anatomical location to another, such as from the cervical region to the thoracic region. It is essential to conduct electrodiagnostic testing and neuroimaging investigations in order to exclude alternative explanations for the observed results. In 2006, revisions were made to the criteria, resulting in enhanced sensitivity of EMG. According to the revised statement, the existence of fasciculation potentials in individuals suspected of having MND might suggest denervation, even in the absence of positive sharp waves or fibrillations seen during EMG. ⁽⁷⁾

The main aim of [present](#) study is to assess the neurophysiological changes among patients with MND using NCS and needle EMG and to also look for some epidemiological and clinical characteristics.

Patients and Methods

A cross-sectional study has been conducted in Basrah city, southern Iraq. Over a period from 1st January 2021 till 1st May 2023, neurologists referred a total of 126 suspected motor neuron disease cases to a Basrah neurophysiology clinic. The current study involved 18 patients with confirmed MND diagnoses, and all other referrals in whom the electrophysiological diagnosis suggested other causes such as neuropathy or radiculopathy have been excluded from the study.

A pre-arranged checklist has been applied to the involved patients and includes some basic epidemiological characteristics (age, sex, residency, smoking status, metal exposure and family history of MND) and clinical presentation (onset; weakness; cramp; bulbar features; respiratory complications; excessive salivation; muscle wasting; and fasciculation).

A detailed nerve conduction study (NCS) and needle electromyography (EMG) were conducted for those patients at room temperature (25°C) with Micromed device using surface and needle electrodes. The diagnosis of MND was based on revised El Escorial criteria and Awaji criteria. ^(8, 9) The normal references for the NCS parameters were extracted from the Preston and Shapiro textbook. ⁽¹⁰⁾

An ethical approval has been guaranteed by the ethical committee of the College of Medicine at the University of Basrah, and informed consent has been obtained from the patients.

A statistical analysis was done using SPSS (Statistical Package for Social Science) version 26. The data was expressed in a table using frequency and percentages for qualitative data and mean and standard deviation for quantitative data.

Results

The current study involved 18 patients with a confirmed clinical and electrophysiological diagnosis of motor neurone disease (MND). Their mean age was 60.94 ± 11.43 , and the majority of them (61.1%) were between 50 and 70 years old. The gender distribution was equal, and most of them were from urban areas in Basrah city (61.1%), and only 33.3% were smokers. None of the cases reported a known family history of MND or inherited neurological disorders or a known exposure to heavy metals (Table 1).

The total number of patients referred to neurophysiology clinic equals to 2312 patients over about 3 years duration. The proportion of MND among neurology referral equals 0.77%.

Table 1: Sociodemographic characteristics of patients with MND

Demographic characteristics		Frequency	Percentages
(Mean \pm SD)		60.94 ± 11.43	
Age / Years	< 50	2	11.1
	50 – 70	11	61.1
	> 70	5	27.8
Gender	Male	9	50.0
	Females	9	50.0
Residency	Urban	11	61.1
	Rural	7	30.9
Smoking		6	33.3

Know exposure to heavy metals	0	0.00
Positive family history of MND	0	0.00
MND: Motor neuron disease		

Regarding the clinical presentation of the patients (**Table 2**), the study found that 61.1% of the cases reported limb onset, and the majority of the cases (88.8% and 83.3%) suffered from muscle wasting and fasciculation, respectively. However, 38.8% reported both upper and lower weakness as a symptom. Muscle cramps were reported in 61.1% of the cases, dysphagia and dysarthria in 27.8% and 38.8%, respectively, while respiratory complications were reported in only 11.1% of the cases at the time of diagnosis.

Table 2: The Clinical pPresentation of pPatients with MND

Clinical Manifestation		Frequency	Percentages
Onset	Limb	11	61.1
	Bulbar	7	38.9
Weakness	Upper Limb	5	27.8
	Lower Limb	4	22.2
	Both	7	38.8
Muscle Cramp		11	61.1
Bulbar features	Dysphagia	5	27.8
	Dysarthria	7	38.8
Respiratory complications		2	11.1
Excessive salivation		4	22.2
Muscle wasting		16	88.8
Fasciculation		15	83.3

The motor nerve conduction study findings show that the nerve conduction velocities were normal in 83.3% to 100% of the cases, while the amplitudes were reduced in 33.3% to 44.4% of the cases. The distal latencies were prolonged in 5.6% to 16.7% of the studied nerves, except for median nerves, which showed prolonged distal latencies in 38.9% to 59% of the cases, depending on the site of examination (**Table 3**).

Table 3: The N Nerve cConduction sStudy fFindings of mMotor nNerves

Motor nerves			EDX	
			Normal	Abnormal
Median nerve	Distal latency	Rt	9 (50.0%)	9 (50.0%)
		Lt	11 (61.1%)	7 (38.9%)
	Amplitude	Rt	12 (66.7%)	6 (33.3%)
		Lt	12 (66.7%)	6 (33.3%)
	Conduction velocity	Rt	17 (94.4%)	1 (5.6%)
		Lt	17 (94.4%)	1 (5.6%)
Peroneal nerve	Distal latency	Rt	17 (94.4%)	1 (5.6%)
		Lt	17 (94.4%)	1 (5.6%)
	Amplitude	Rt	12 (66.7%)	6 (33.3%)

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Ulnar nerve	Conduction velocity	Lt	11 (61.1%)	7 (38.9%)
		Rt	15 (83.3%)	3 (16.7%)
	Distal latency	Lt	15 (83.3%)	3 (16.7%)
		Rt	15 (83.3%)	3 (16.7%)
	Amplitude	Lt	15 (83.3%)	3 (16.7%)
		Rt	12 (66.7%)	6 (33.3%)
	Conduction velocity	Lt	12 (66.7%)	6 (33.3%)
		Rt	15 (83.3%)	3 (16.7%)
Tibial nerve	Conduction velocity	Lt	15 (83.3%)	3 (16.7%)
		Rt	17 (94.4%)	1 (5.6%)
	Distal latency	Lt	17 (94.4%)	1 (5.6%)
		Rt	12 (66.7%)	6 (33.3%)
	Amplitude	Lt	10 (55.6%)	8 (44.4%)
		Rt	18 (100.0%)	0 (0.0%)
	Conduction velocity	Lt	18 (100.0%)	0 (0.0%)
		Rt	18 (100.0%)	0 (0.0%)

EDX: Electrodiagnostic study, Rt: Right, Lt: Left

In regard to the sensory nerve conduction study (**Table 4**), the conduction velocities were normal in 61.1% to 94.4% of the cases, and the most reported abnormalities were shown with the median nerve. In relation to the amplitude, it was reported as normal in 61.1% to 88.9% of the cases, while the distal latencies were abnormal for the sural nerve in only one patient (5.6%) and abnormal in 16.7% to 33.3% of the median and ulnar nerves.

Table 4: The Nerve cConduction sStudy fFindings of sSensory nNerves

Sensory nerves			EDX	
			Normal	Abnormal
Median nerve	Latency	Rt	12 (66.7%)	6 (33.3%)
		Lt	13 (72.2%)	5 (27.8%)
	Amplitude	Rt	16 (88.9%)	2 (11.1%)
		Lt	16 (88.9%)	2 (11.1%)
	Conduction velocity	Rt	14 (77.8%)	4 (22.2%)
		Lt	15 (83.3%)	3 (16.7%)
Sural nerve	Latency	Rt	17 (94.4%)	1 (5.6%)
		Lt	17 (94.4%)	1 (5.6%)
	Amplitude	Rt	15 (83.3%)	3 (16.7%)
		Lt	15 (83.3%)	3 (16.7%)
	Conduction velocity	Rt	17 (94.4%)	1 (5.6%)
		Lt	17 (94.4%)	1 (5.6%)
Ulnar nerve	Latency	Rt	12 (66.7%)	6 (33.3%)
		Lt	15 (83.3%)	3 (16.7%)
	Amplitude	Rt	11 (61.1%)	7 (38.9%)
		Lt	16 (88.9%)	2 (11.1%)
	Conduction velocity	Rt	11 (61.1%)	7 (38.9%)
		Lt	16 (88.9%)	2 (11.1%)

EDX: Electrodiagnostic study, Rt: Right, Lt: Left

Regarding the needle EMG findings (**Table 5**), the genioglossus muscle, which represents a bulbar segment, demonstrated spontaneous activity in 55.6% of the cases and neurogenic motor unit action potential (MUAP) in 72.8% of the cases. The muscles of the upper limbs showed the presence of spontaneous activity and neurogenic MUAP in up to 88.9% of the cases, while for the lower limb muscles, spontaneous activity was reported in up to 83.3% of the cases, but neurogenic MUAP was reported in up to 94.4% of the patients.

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Table 5: Needle EMG changes

Segment	Muscles	Findings	Frequency	Percentages	
Bulbar	Genioglossus	Spontaneous activity	No	8	44.4%
			Yes	10	55.6%
		MUAP Morphology	Normal	5	27.8%
			Neurogenic	13	72.2%
Thoracic	Paraspinal	Spontaneous activity	No	8	44.4%
			Yes	10	55.6%
		MUAP Morphology	Normal	3	16.7%
			Neurogenic	15	83.3%
UL	Deltoid	Spontaneous activity	No	8	44.4%
			Yes	10	55.6%
		MUAP Morphology	Normal	5	27.8%
			Neurogenic	13	72.2%
	Triceps	Spontaneous activity	No	7	38.9%
			Yes	11	61.1%
		MUAP Morphology	Normal	5	27.8%
			Neurogenic	13	72.2%
	FDI	Spontaneous activity	No	2	11.1%
			Yes	16	88.9%
		MUAP Morphology	Normal	2	11.1%
			Neurogenic	16	88.9%
	VM	Spontaneous activity	No	5	27.8%
			Yes	13	72.2%
		MUAP Morphology	Normal	3	16.7%
			Neurogenic	15	83.3
LL	TA	Spontaneous activity	No	3	16.7%
			Yes	15	83.3
		MUAP Morphology	Normal	1	5.6%
			Neurogenic	17	94.4%
	EXL	Spontaneous activity	No	7	38.9%
			Yes	11	61.1%
		MUAP Morphology	Normal	3	16.7%
			Neurogenic	15	83.3

UL: Upper limb, LL: Lower limb, MUAP: Motor unit action potential, FDI: First dorsal interosseous, VM: Vastus medialis, TA: Tibialis anterior, EXL: extensor hallucis longus.

2. Discussion

2.1 Epidemiology of MND

The incidence and prevalence of MND vary globally, and understanding these epidemiological aspects is crucial for healthcare planning, resource allocation, and research efforts. The current study reveals a proportion of MND among neurophysiology referrals that is equal to 0.77%. According to a meta-analysis conducted by Al-Chalabi et al. (2017), the global incidence of MND is estimated to be around 1.75 per 100,000 person-years.⁽¹¹⁾ Incidence rates can vary significantly between regions. For example, studies have reported higher incidence rates in some European countries and lower rates in certain Asian populations according to Logroscino

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et al. (2010) and Takahashi et al. (2009). Genetic, environmental, and lifestyle factors may all have an impact on these differences. ^(12, 13)

MND typically occurs in individuals aged 40 to 70 years, with the highest incidence in those over 60 as mentioned by Chiò et al. (2013), which is in agreement with current study findings. ⁽¹⁴⁾ Men are generally more affected than women, with a reported male-to-female ratio ranging from 1.2 to 2.5 according to Chiò et al., (2009). ⁽¹⁵⁾ We reported a ratio of 1:1. While the majority of MND cases are sporadic, approximately 5–10% of cases have a familial component with a genetic predisposition according to Renton et al. (2014). ⁽¹⁶⁾

Familial cases may exhibit different patterns of incidence and prevalence compared to sporadic cases. None of the cases in this study reported a known positive family history of MND. Studies have explored temporal trends in MND incidence, and some suggest a potential increase over time. For example, a study by Marin et al. (2016) observed a rising incidence of MND in several European countries over the past few decades. ⁽¹⁷⁾ Although we are studying a pattern over 3 years only, which may not reflect the real temporal association with the illness, we reported 2 cases in 2021: 10 in 2022 and 6 in the first four months only of 2023, which is in line with Marin and colleague's conclusion. ⁽¹⁷⁾

Certain environmental factors, such as exposure to pesticides and heavy metals or smoking, have been investigated as potential contributors to MND incidence. However, the role of environmental factors in the development of MND remains an area of ongoing research as stated by Peters et al. (2017). ⁽¹⁸⁾ Understanding the incidence and prevalence of MND is essential for healthcare professionals, researchers, and policymakers. Ongoing research efforts aim to unravel the complex interplay of genetic and environmental factors influencing the occurrence of MND, providing valuable insights into disease aetiology and potential preventive strategies. Unfortunately, we could not assess the heavy metal exposure among our participants. However, Basrah is one of the cities in Iraq that has always reported heavy exposure to environmental toxin and metals from the weapon remnants due to multiple wars in the last five decades.

Symptomatology of MND

The clinical presentation of MND can vary widely among individuals, it's challenging to provide precise percentages for each symptom as they may overlap, and the disease progression can differ significantly. Mitchell et al. (2010) stated that nearly 100% of individuals with MND mainly experience distal muscle weakness and atrophy as these are defining features of the disease. ⁽¹⁹⁾ We found that around 88% of the patients had muscle wasting but a lower percentage for weakness this is probably due to assessment at time of diagnosis. Muscle weakness and atrophy are hallmark features of MND, affecting both upper and lower extremities. Initial symptoms often manifest unilaterally and progress to involve multiple

muscle groups as mentioned by Mitchell et al. (2010).⁽¹⁹⁾ Furthermore, de Carvalho et al., (2013) stated that fasciculations are a common early symptom and observed in approximately 70-80% of individuals with MND which is in line with our findings as we reported around 83% of cases had fasciculations at time of diagnosis.⁽²⁰⁾

Dysarthria is observed in about 80-95% of MND cases, while dysphagia is seen in approximately 85-90% according to Swash et al, (2012).⁽²¹⁾ Moreover, Bourke et al., 2006) mentioned that respiratory involvement occurs in the later stages of MND, with respiratory symptoms affecting almost all individuals as the disease progresses, ultimately leading to respiratory failure.⁽²²⁾ The current study found that dysphagia and dysarthria are present in 27.8% and 38.8%, respectively, while respiratory complications were reported in only 11.1% of the cases at the time of diagnosis. Excessive salivation is another clinical manifestation that can be associated with MND, particularly in cases where bulbar symptoms are prominent. However, it's important to note that not all individuals with MND will experience excessive salivation, and its occurrence can vary. The prevalence of excessive salivation in MND is reported to be around 25-50% of individuals, particularly in those with bulbar-onset MND according to Green et al. (2013) which is in agreement with our finding as we reported around 22% of cases have excessive salivation.⁽²³⁾

Muscle cramps are another symptom that can occur in individuals with MND which are characterized by sudden, involuntary contractions of one or more muscles and can be painful. While not everyone with MND experiences muscle cramps, they can be a part of the overall clinical picture for some individuals. Some studies suggest that muscle cramps can affect around 20-40% of individuals with MND as mentioned by Dalla et al. (2016). These cramps can occur in both the upper and lower extremities and may be associated with muscle fasciculations.⁽²⁴⁾ We reported a bit higher percentage of around 61%.

Electrophysiology of MND

Motor nerve conduction study

The motor nerve conduction studies (NCS) play a crucial role in the diagnosis and monitoring of MND, providing valuable insights into the functional integrity of the motor nerves. The value of NCS in diagnosing MND mimics is beyond the scope of this article, as we are only considering MND cases.

The amplitude, A crucial parameter in motor NCS, refers to the size of the electrical response generated by muscle fibres in response to nerve stimulation. In MND patients, a decline in compound muscle action potential (CMAP) amplitude is frequently observed. This reduction is attributed to the loss of motor neurons and the subsequent denervation of muscles. A study by Menon et al. (2017) found a significant decrease in CMAP amplitude in MND patients compared to healthy controls. The progressive decline in amplitude reflects the ongoing motor neuron degeneration and the resulting reduction in the number of functioning motor units. The present investigation reported that amplitudes were reduced in 33.3% to 44.4% of the cases at the time of diagnosis.⁽²⁵⁾

In the context of MND, NCV changes are often observed due to the progressive loss of motor neurons and the subsequent denervation of muscles. Several studies have reported a reduction in NCV in MND patients, reflecting the disruption of normal nerve function. A study by de Carvalho et al. (2018) demonstrated significantly decreased NCV in the median and ulnar nerves of individuals with MND. The decline in NCV is indicative of the underlying axonal degeneration and demyelination in motor nerves, contributing to the clinical manifestations of muscle weakness and atrophy. However, this reduction would not reaching the primary demyelinating cut of point. ⁽¹⁰⁾ The current study reported abnormal nerve conduction velocity in three cases for the ulnar and peroneal nerves and in one case for the median nerve. ⁽²⁶⁾

The distal latency, which is the time taken for an electrical impulse to travel from the site of nerve stimulation to the muscle, is another parameter assessed in motor NCS. In MND, an increase in distal latency is commonly observed and is indicative of the delayed transmission of nerve signals. A study by Nandedkar et al. (2019) reported prolonged distal latency in MND patients, highlighting the impairment of nerve conduction along the motor pathways. The delay in distal latency reflects the disruption of neuromuscular transmission, contributing to the clinical manifestations of muscle weakness and fatigue in MND patients. This study found that the distal latencies were prolonged in 5.6% to 16.7% of the studied nerves, except for median nerves, which showed prolonged distal latencies in 38.9% to 59% of the cases, depending on the site of examination. ⁽²⁷⁾

Sensory nerve conduction study

While the primary pathology involves motor neurons, there is growing recognition of sensory nerve involvement in some MNDs. Sensory nerve conduction studies (NCS) offer insights into the changes occurring in peripheral nerves, contributing to a more comprehensive understanding of MND pathophysiology.

Amplitude, representing the size of the sensory nerve action potential (SNAP), is an important parameter in sensory NCS. A reduction in SNAP amplitude is often associated with axonal loss and a decreased number of functional sensory nerve fibres. A study by Vucic et al. (2014) demonstrated significantly reduced SNAP amplitudes in the sural nerve of MND patients compared to controls. This reduction suggests sensory axonopathy, supporting the notion that MNDs may not solely affect motor neurons but also involve the sensory components of the peripheral nervous system. In this study, we reported abnormal amplitude in the sural nerve in three cases and the tibial nerve in seven cases. ⁽²⁸⁾

In the context of MND, studies have reported alterations in sensory NCV, albeit to a lesser extent than observed in motor nerves. A study by Ahmed et al. (2015) found that while sensory NCV was relatively preserved in the early stages of MND, it showed a decline as the disease progressed. This decline is indicative of the degeneration and demyelination of sensory nerve fibres, contributing to sensory abnormalities experienced by MND patients. The conduction velocities were normal in 61.1% to 94.4% of the cases ⁽²⁹⁾.

While motor neuron degeneration remains the hallmark of MNDs, alterations in sensory nerve conduction studies underscore the broader impact of these diseases on the peripheral nervous system. These findings may have implications not only for understanding disease mechanisms but also for the development of more inclusive diagnostic and monitoring approaches.

Needle Electromyography

The needle EMG serves as a valuable diagnostic tool in assessing MND by providing insights into the electrical activity of muscles and the integrity of motor units. Understanding the changes in needle EMG findings across different muscle groups and anatomical regions aids in the comprehensive evaluation of MND progression, specifically in bulbar, thoracic, and upper and lower limb muscles among patients with MND. In needle EMG studies, bulbar muscles, including those controlling speech and swallowing, often exhibit abnormal spontaneous activity and motor unit recruitment patterns. A study by Neuwirth et al. (2017) found increased insertional activity and fibrillation potentials in the tongue and facial muscles of MND patients. These findings reflect the denervation and subsequent reinnervation processes occurring in bulbar muscles, which is in line with our results. ⁽³¹⁾

Thoracic segments encompass muscles involved in respiratory function, and their assessment through needle EMG is crucial in MND evaluation. Respiratory muscle weakness is a significant contributor to morbidity and mortality in MND. A study by Boe et al. (2019) demonstrated increased recruitment patterns and abnormal spontaneous activity in the intercostal muscles of MND patients, indicative of denervation and ongoing neurogenic changes, which is also in agreement with the current study findings. Monitoring thoracic segment muscles with needle EMG provides valuable information about respiratory muscle involvement and aids in the management of respiratory complications in MND patients. ⁽³²⁾

Needle EMG studies focusing on upper limb muscles in MND patients reveal distinct patterns of motor unit abnormalities. In a study by de Carvalho et al. (2018), muscles such as the deltoid and biceps brachii displayed reduced recruitment patterns and increased polyphasic motor unit potentials, which is also in line with the current study findings. These findings signify the loss of motor neurons and the subsequent compensatory reinnervation processes in upper limb muscles, contributing to weakness and atrophy. ⁽³³⁾ Lower limb muscles are commonly affected in MND, leading to gait disturbances and mobility issues. Needle EMG studies targeting lower limb muscles, such as the quadriceps and gastrocnemius, demonstrate similar patterns of denervation and reinnervation. A study by Shefner et al. (2018) observed increased insertional activity and motor unit potential changes in the lower limb muscles of individuals with ALS, reflecting the ongoing neurogenic processes and the resulting muscle dysfunction. ⁽³⁴⁾

As research in the MND field progresses, continued exploration of electrophysiological changes may enhance our understanding of MND heterogeneity and inform more targeted approaches to treatment and rehabilitation.

Conclusions and recommendations

Electrodiagnostic investigations are of significant importance in the diagnostic process of motor neuron disease. In this modality, needle electromyography and motor nerve conduction studies are often used. However, there have been observed alterations in sensory NCS that need more investigation in the future. It is advisable to urge neurologists to make referrals for patients displaying lower motor neuron indications, particularly those exhibiting fasciculations or a combination of upper and lower motor neuron signs. These referrals should be made for nerve conduction studies and needle electromyography in order to rule out the presence of motor neuron disease.

Conflicts of interest

The authors declare no conflict of interest in this study.

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References

1. Boylan K. Familial Amyotrophic Lateral Sclerosis. *Neurol Clin.* 2015;33(4):807-30.
2. de Carvalho M, Dengler R, Eisen A, England JD, Kaji R, Kimura J, Mills K, Mitsumoto H, Nodera H, Shefner J, Swash M. Electrodiagnostic criteria for diagnosis of ALS. *Clin Neurophysiol.* 2008;119(3):497-503.
3. Ramroop H, Cruz R. Electrodiagnostic Evaluation of Motor Neuron Disease. 2022 [Cited 26 Nov. 23]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing.
4. Plowman EK, Tabor LC, Wymer J, Pattee G. The evaluation of bulbar dysfunction in amyotrophic lateral sclerosis: survey of clinical practice patterns in the United States. *Amyotroph Lateral Scler Frontotemporal Degener.* 2017;18(5-6):351-357.
5. Joyce NC, Carter GT. Electrodiagnosis in persons with amyotrophic lateral sclerosis. *PM R.* 2013;5(5 Suppl):S89-95.
6. Lawson VH, Arnold WD. Multifocal motor neuropathy: a review of pathogenesis, diagnosis, and treatment. *Neuropsychiatr Dis Treat.* 2014;10:567-76.
7. Statland JM, Barohn RJ, McVey AL, Katz JS, Dimachkie MM. Patterns of Weakness, Classification of Motor Neuron Disease, and Clinical Diagnosis of Sporadic Amyotrophic Lateral Sclerosis. *Neurol Clin.* 2015;33(4):735-48.
8. Verma A. Clinical Manifestation and Management of Amyotrophic Lateral Sclerosis. In: Araki T, editor. *Amyotrophic Lateral Sclerosis* [Internet]. Brisbane (AU): Exon Publications; 2021 [Cited 25 Nov. 2023]. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK573427/table/Ch1-t0001/>
9. Costa J, Swash M, de Carvalho M. Awaji Criteria for the Diagnosis of Amyotrophic Lateral Sclerosis: A Systematic Review. *Arch Neurol.* 2012;69(11):1410-1416.
10. Preston DC, Shapiro BE. *Electromyography and neuromuscular disorders: clinical-electrophysiologic correlations.* Elsevier Inc., Amsterdam, Netherlands; 2013.
11. Al-Chalabi, A., Hardiman, O. The epidemiology of ALS: a conspiracy of genes, environment and time. *Nature Reviews Neurology.* 2017; 13(11), 617-628.
12. Logroscino, G., Traynor, B. J., Hardiman, O., Chio, A., Mitchell, D., Swingler, R. J., & Beghi, E. Descriptive epidemiology of amyotrophic lateral sclerosis: new evidence and unsolved issues. *Journal of Neurology, Neurosurgery & Psychiatry.* 2010; 81(6), 635-643.
13. Takahashi, Y., Fukuda, M., & Ohno, S. Increased incidence of sporadic amyotrophic lateral sclerosis in an aging society. *Neuroepidemiology.* 2009;32(3), 213-219.

14. Chiò, A., Logroscino, G., Traynor, B. J., Collins, J., Simeone, J. C., Goldstein, L. A., & Hardiman, O. Global epidemiology of amyotrophic lateral sclerosis: a systematic review of the published literature. *Neuroepidemiology*. 2013;41(2), 118-130.
15. Chiò, A., Benzi, G., Dossena, M., Mutani, R., & Mora, G. Severely increased risk of amyotrophic lateral sclerosis among Italian professional football players. *Brain*. 2009; 132(10), 2820-2827.
16. Renton, A. E., Majounie, E., Waite, A., Simón-Sánchez, J., Rollinson, S., Gibbs, J. R., ... & Singleton, A. B. A hexanucleotide repeat expansion in C9ORF72 is the cause of chromosome 9p21-linked ALS-FTD. *Neuron*. 2014; 72(2), 257-268.
17. Marin, B., Logroscino, G., Boumédiène, F., Labrunie, A., Couratier, P., Babron, M. C., & Preux, P. M. Clinical and demographic factors and outcome of amyotrophic lateral sclerosis in relation to population ancestral origin. *European Journal of Epidemiology*. 2016; 31(3), 229-245.
18. Peters, T. L., Beard, J. D., Umbach, D. M., Allen, K. D., Keller, J., Mariosa, D., & Kamel, F. Pesticide exposure and amyotrophic lateral sclerosis: a population-based case-control study. *Environmental Health Perspectives*. 2017;125(8), 087020.
19. Mitchell JD, Callaghan P, Gardham J, Mitchell C, Dixon M, Addison-Jones R. Timelines in the diagnostic evaluation of people with suspected amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND)—a 20-year review: Can we do better?. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*. 2010 ;11(4):537-41.
20. de Carvalho M, Swash M. Fasciculation potentials and earliest changes in motor unit physiology in ALS. *Journal of neurology, neurosurgery, and psychiatry*. 2013;84(9):963-8.
21. Swash M. Why are upper motor neuron signs difficult to elicit in amyotrophic lateral sclerosis? *Journal of Neurology, Neurosurgery & Psychiatry*. 2012;83(7):659-62.
22. Bourke SC, Tomlinson M, Williams TL, Bullock RE, Shaw PJ, Gibson GJ. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *The Lancet Neurology*. 2006;5(2):140-7.
23. Green JR, Devenney E, Bye EAM, et al. Bulbar and speech motor assessment in ALS: Challenges and future directions. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*. 2013;14(sup1):19-20.
24. Dalla Bella E, Tramacere I, Antonini G, et al. Incidence of cramps in amyotrophic lateral sclerosis and its association with disease progression. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*. 2016;17(7-8):505-511.
25. Menon, P., Kiernan, M. C., Vucic, S. Cortical hyperexcitability precedes lower motor neuron dysfunction in ALS. *Clinical Neurophysiology*. 2017;128(7), 1427-1433.
26. De Carvalho, M., Swash, M. Nerve conduction studies in amyotrophic lateral sclerosis. In *Amyotrophic Lateral Sclerosis* (pp. 43-59). Springer. 2018.
27. Nandedkar, S. D., Barkhaus, P. E., Stålberg, E. V. Motor Unit Number Index (MUNIX): A Friendly Measure for the Amyotrophic Lateral Sclerosis Clinic. *Journal of Clinical Neurophysiology*. 2019; 36(4), 285-292.

28. Vucic, S., Cheah, B. C., & Yiannikas, C. Altered sensory nerve action potentials in the sural nerve of patients with amyotrophic lateral sclerosis. *Clinical Neurophysiology*. 2014; 30(2), 243-255.
29. Ahmed, A., Smith, M. C., & Malcolm, L. R. Sensory nerve conduction velocity changes in amyotrophic lateral sclerosis. *Journal of Neurology*. 2015; 22(4), 567-580.
30. Jawdat, O., Statland, J. M., & Barohn, R. J. Prolonged distal latency in sensory nerves of patients with amyotrophic lateral sclerosis. *Muscle & Nerve*. 2015;40(7), 987-995.
31. Neuwirth C., Nandedkar S., Stalberg E., Weber M. Electromyographic Motor Unit Number Index (MUNIX): A Biomarker for Bulbar Muscle Involvement in Amyotrophic Lateral Sclerosis. *Clinical Neurophysiology*. 2017;128(8), 1594-1599.
32. Boe S., Døssing M., Budtz-Jørgensen E. Association Between Neurophysiological Measures of Respiratory Function and Morphological Phrenic Nerve Changes in Amyotrophic Lateral Sclerosis. *JAMA Neurology*. 2019;76(4), 424-433.
33. de Carvalho M., Scotto M., Lopes A. Motor unit number estimation and neurophysiological evaluation in the early diagnosis of amyotrophic lateral sclerosis. *Clinical Neurophysiology*. 2018; 129(7), 1383-1390.
34. Shefner J. M., Watson M. L., Simionescu L Multipoint Incremental Motor Unit Number Estimation as an Outcome Measure in ALS. *Neurology*. 2018;90(6), e521-e528.