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Original Research Article

# Clinical and Demographic Patterns in Motor Neuron Disease: An Observational Study from a Tertiary Care Centre in Northeast India

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**Abstract:**

**Background:** Motor neuron disease (MND) encompasses a group of progressive neurodegenerative disorders affecting motor neurons. Understanding the clinical and demographic patterns of MND in specific populations is crucial for improving diagnosis and management. This study aimed to analyze the clinical and demographic characteristics of MND patients presenting to a tertiary care center in Northeast India.

**Methods:** This observational study analyzed 40 patients diagnosed with MND at the Department of Neurology, Gauhati Medical College, and Guwahati. Data collected included demographics, clinical presentation, El Escorial classification, and one-year survival outcomes. Statistical analysis was performed using appropriate tests with significance set at $p<0.05$.

**Results:** The study cohort (n=40) showed male predominance (70%) with a mean age of 58.8 years. Spinal onset was most common (50%), followed by bulbar (30%), generalized (10%), and bibrachial (10%) onset. Bulbar involvement was present in 72.5% of patients. The majority were classified as probable ALS (62.5%) by El Escorial criteria. One-year survival was 82.5%. Numerical differences were observed in survival rates across onset types, with bibrachial onset showing 100% survival and generalized onset showing 50% survival, but these did not reach statistical significance ($p=0.276$). A trend toward lower survival in patients with definite ALS was noted, potentially reflecting more advanced disease at presentation.

**Conclusions:** This study reveals distinct clinical patterns of MND in Northeast India, with high prevalence of bulbar involvement and relatively favorable one-year survival rates. The heterogeneity in presentation and outcomes based on onset type highlights the importance of individualized prognostic assessment and management strategies.

**Keywords:** Motor Neuron Disease, Amyotrophic Lateral Sclerosis, Northeast India, El Escorial Criteria, Survival Analysis.



## Introduction

Motor neuron disease (MND) represents a heterogeneous group of progressive neurodegenerative disorders characterized by selective loss of motor neurons in the brain and spinal cord.[1] The global incidence of MND ranges from 0.6 to 3.8 per 100,000 person-years, with significant geographical and ethnic variations.[2] Amyotrophic lateral sclerosis (ALS), the most common form of MND, typically presents between the fifth and seventh decades of life with a male predominance.[3]

The clinical phenotype of MND is highly variable, with patients presenting with spinal, bulbar, or generalized onset patterns.[4] This heterogeneity poses diagnostic challenges and influences prognostic assessment. The revised El Escorial criteria remain the standard for diagnostic classification, categorizing patients into definite, probable, and possible ALS based on clinical and

electrophysiological findings.[5] In India, epidemiological data on MND remains limited, particularly from the northeastern region. [6] Understanding regional patterns of disease presentation and outcomes is essential for optimizing patient care and resource allocation. This study aimed to characterize the clinical and demographic patterns of MND patients presenting to a tertiary care center in Northeast India.

## Materials and Methods

**Study Design and Setting:** This observational, cross-sectional study was conducted at the Department of Neurology, Gauhati Medical College and Hospital, Guwahati, Assam. Data was collected from patients diagnosed with MND between January 2022 and December 2023.

**Study Population:** Consecutive patients diagnosed with MND according to the revised El Escorial criteria were included. Exclusion criteria included patients with incomplete clinical data, those lost to follow-up before one year, and patients with other neurodegenerative disorders mimicking MND.

**Data Collection:** A structured proforma was used to collect demographic data (age, sex), clinical characteristics (duration of symptoms, onset type, bulbar involvement), presence of frontotemporal dementia (FTD), family history, comorbidities, El Escorial classification, and one-year survival status.

All patients underwent comprehensive neurological examination, electromyography, and nerve conduction studies.

**Statistical Analysis:** Data was analyzed using appropriate statistical software. Descriptive statistics were presented as mean ± standard deviation for continuous variables and frequencies with percentages for categorical variables. Subgroup analyses were performed to examine associations between onset type, El Escorial classification, and survival outcomes.

Chi-square test was used for categorical variables, with $p<0.05$ considered statistically significant.

**Ethical Considerations:** The study was approved by the Institutional Ethics Committee. Written informed consent was obtained from all participants or their legal representatives.

## Results

**Demographic Characteristics:** The study included 40 patients with confirmed MND. Table 1 summarizes the demographic and clinical characteristics of the cohort.

**Table 1: Demographic and Clinical Characteristics of MND Patients (n=40)**

| Characteristic | | n (%) |
|---|---|---|
| Age (years) | Mean ± SD | 58.8 ± 9.5 |
| | Range | 36-75 |
| Sex | Male | 28 (70.0) |
| | Female | 12 (30.0) |
| | Male: Female ratio | 2.3:1 |
| Age Distribution | 30-39 years | 4 (10.0) |
| | 40-49 years | 3 (7.5) |
| | 50-59 years | 10 (25.0) |
| | 60-69 years | 17 (42.5) |
| | 70-79 years | 6 (15.0) |
| Duration of symptoms (months) | Mean ± SD | 12.3 ± 5.2 |
| | Range | 6 to 13 |

The cohort demonstrated male predominance with a male-to-female ratio of 2.3:1. The peak incidence occurred in the seventh decade (60-69 years), accounting for 42.5% of cases. Notably, females presented at an older age (mean 65.1 years) compared to males (mean 56.1 years).

## Clinical Patterns

**Table 2: Clinical Characteristics and Disease Patterns**

| Characteristic | | n (%) |
|---|---|---|
| Onset Type | Spinal | 20 (50.0) |
| | Bulbar | 12 (30.0) |
| | Generalized | 4 (10.0) |
| | Bibrachial | 4 (10.0) |
| Bulbar Involvement | Present | 29 (72.5) |
| | Absent | 11 (27.5) |
| Frontotemporal Dementia | Present | 3 (7.5) |
| | Absent | 37 (92.5) |
| Family History | Positive | 2 (5.0) |
| | Negative | 38 (95.0) |
| Comorbidities | Present | 22 (55.0) |
| | Absent | 18 (45.0) |

Spinal onset was the most common presentation (50%), followed by bulbar onset (30%). The high prevalence of bulbar involvement (72.5%) across all onset types is noteworthy. Among comorbidities, hypertension was most frequent (n=10), followed by type 2 diabetes mellitus (n=4).

**El Escorial Classification:** The distribution according to El Escorial criteria revealed: probable ALS (62.5%, n=25), definite ALS (22.5%, n=9), and possible ALS (15.0%, n=6).

**Subgroup Analysis**

**Onset Type versus El Escorial Classification** : Analysis revealed distinct patterns in diagnostic certainty across onset types. All patients with generalized onset (100%) met criteria for definite ALS, while bulbar onset predominantly presented as probable ALS (83.3%).

Spinal onset showed the most heterogeneous distribution: probable (70%), possible (20%), and definite (10%). Bibrachial onset displayed equal distribution between definite and probable (25% each), with 50% classified as possible ALS.

**Survival Analysis**: One-year survival analysis demonstrated significant variations across onset types (Table 3).

**Table 3: One-Year Survival by Onset Type**

| Onset Type | Total (n) | Alive n (%) | Deceased n (%) |
|---|---|---|---|
| Bibrachial | 4 | 4 (100.0) | 0 (0.0) |
| Spinal | 20 | 17 (85.0) | 3 (15.0) |
| Bulbar | 12 | 10 (83.3) | 2 (16.7) |
| Generalized | 4 | 2 (50.0) | 2 (50.0) |
| Overall | 40 | 33 (82.5) | 7 (17.5) |

Chi-square test: $\chi^2 = 3.867$, $p = 0.276$ (not significant)

Bibrachial onset demonstrated the best prognosis with 100% one-year survival, while generalized onset showed the poorest outcome with 50% mortality. However, these differences did not reach statistical significance (p=0.276).

**El Escorial Classification versus Survival:** A trend toward lower survival was observed with increasing diagnostic certainty. Patients classified as possible ALS showed 100% one-year survival (6/6), probable ALS had 84% survival (21/25), while definite ALS demonstrated 66.7% survival (6/9). This trend, while numerically notable, was not subjected to statistical testing due to small subgroup sizes and may reflect more advanced disease in patients meeting definite criteria.

**Bulbar Involvement by Onset Type**: As expected, all patients with bulbar onset (100%) and generalized onset (100%) had bulbar involvement. Spinal onset showed bulbar involvement in 55% of cases, while bibrachial onset had 50% bulbar involvement.

**Age-Related Patterns:** Mean age varied by onset type: bibrachial (51.5 years), spinal (55.1 years), bulbar (65.1 years), and generalized (65.5 years). This trend toward younger age in limb-onset variants compared to bulbar and generalized forms aligns with established patterns.[7]

**Bulbar Involvement and Outcomes:** While bulbar involvement was present in 72.5% of our cohort, it did not significantly affect one-year survival (p=0.69). This finding may reflect the short follow-up duration, as bulbar involvement is a well-established predictor of poor prognosis in longer-term studies.

**Discussion**

Our study provides comprehensive data on MND patterns from Northeast India, addressing a

significant gap in regional epidemiological information. We found several notable patterns that both align with and diverge from international literature. The male predominance (70%) and male-to-female ratio of 2.3:1 observed in our cohort is consistent with global trends, where ratios typically range from 1.5-2.5:1.[8] However, the mean age at presentation (58.8 years) is slightly younger than Western populations, where peak incidence occurs in the seventh to eighth decade.[9] This earlier presentation may reflect genetic, environmental, or healthcare access factors specific to our population. The distribution of onset types, with spinal predominance (50%) followed by bulbar (30%), mirrors patterns reported from other Asian cohorts.[10] However, the high overall prevalence of bulbar involvement (72.5%) across all onset types is noteworthy and exceeds rates typically reported in Western literature (25-30%).[11] While bulbar involvement is a known predictor of poor prognosis in long-term studies, in our cohort it did not significantly affect one-year mortality (p=0.69), possibly due to the short follow-up duration. Our one-year survival rate of 82.5% is relatively favorable compared to historical cohorts, where median survival from symptom onset typically ranges from 2-4 years.[12] While we observed numerical differences in survival across onset types, with bibrachial onset showing 100% survival versus 50% in generalized onset, these differences did not reach statistical significance (p=0.276). The superior outcome in bibrachial onset may reflect the more restricted nature of motor neuron involvement in this variant.[13] The trend toward lower survival with increasing El Escorial diagnostic certainty represents an intriguing finding. Patients meeting criteria for definite ALS had poorer survival (66.7%) compared to those with possible ALS (100%). This observation likely reflects lead-time bias, as patients with more widespread clinical signs meeting definite criteria are inherently at a more advanced disease stage.[14] This finding emphasizes the limitations of current diagnostic criteria in early disease detection and prognostication. The low prevalence of FTD (7.5%) and positive family history (5%) in our cohort aligns with reported rates in sporadic ALS populations.[15] The presence of comorbidities in 55% of patients, predominantly vascular risk factors, highlights the importance of comprehensive medical management in this population.

**Clinical Implications:** Several findings from this study have direct clinical relevance. The high prevalence of bulbar involvement necessitates early and regular assessment of bulbar function, even in patients with limb-onset disease. The favorable one-year survival rates suggest that aggressive supportive care may positively impact short-term outcomes in our setting. The phenotypic heterogeneity observed reinforces the need for individualized management approaches based on onset type and progression patterns.

**Study Limitations:** This study has several limitations. The single-center design may limit generalizability to other regions of Northeast India. The relatively small sample size, particularly for less common onset types, limits statistical power for some subgroup analyses. The one-year follow-up period, while providing valuable short-term outcome data, does not capture the full disease trajectory. Additionally, we did not perform genetic testing, which may have provided insights into familial cases and genotype-phenotype correlations.

## Conclusions

Our study provides the first comprehensive analysis of MND patterns from Northeast India, revealing distinct demographic and clinical characteristics. The high prevalence of bulbar involvement, numerical differences in survival patterns based on onset type, and the trend toward lower survival with increasing diagnostic certainty highlight the complex nature of MND in our population. While survival differences across onset types did not reach statistical significance in our cohort, these patterns warrant further investigation in larger studies. These findings underscore the need for early diagnosis, phenotype-specific prognostication, and comprehensive multidisciplinary care. Future multicenter studies with longer follow-up periods and genetic characterization are warranted to further elucidate MND patterns in this region.

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