An Ambispective Study on Clinical Profile, Types of ALS, and Functional Outcomes of ALS Patients in a Tertiary Care Centre

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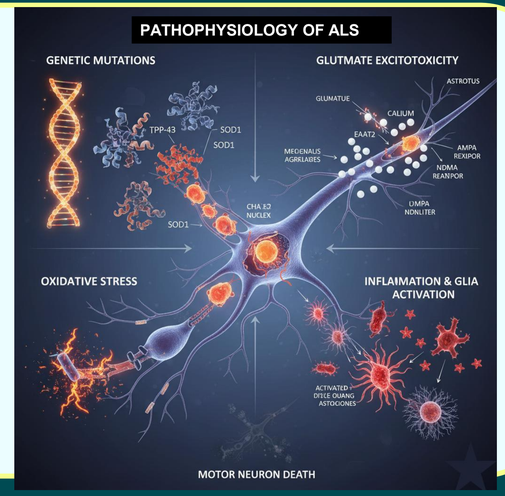
INTRODUCTION:

Amyotrophic lateral sclerosis (ALS) is a progressive, fatal neurodegenerative disease affecting the upper and lower motor neurons, causing significant morbidity and eventual mortality. The hallmark clinical feature is a combination of upper motor neuron signs (spasticity, hyperreflexia) and lower motor neuron signs (muscle weakness, atrophy, fasciculations). Classified into Sporadic ALS (90-95% cases), and familial ALS (5-10%). Clinically, subdivided as Bulbar-onset, Limb-onset, Respiratory-onset, Limb-bulbar, pseudo-bulbar, and other overlap subtypes. The prevalence in India is reported to be around 5 per 100,000, with a younger age of onset and a longer symptom duration compared to Western populations.



AIMS:

To evaluate the clinical characteristics, subtype distribution, diagnostic findings, and functional outcomes of ALS patients over a 2.5-year period at a tertiary care center in South India.

MATERIALS AND METHODS:

This ambispective observational study included 49 ALS patients diagnosed between January 2023 and April 2025.

Medical records were analyzed for demographics, clinical features, neurophysiological findings, imaging results, and diagnostic classification based on revised El Escorial and Gold Coast criteria. Functional outcomes were assessed using ALSFRS-R-SE scores at baseline and a prospective follow-up.

Inclusion criteria: ALS patients who presented to the department of neurology with age >18 yrs of age .

Exclusion criteria: Diseases like vitamin B12 deficiency , cervical and lumbar myelopathy, hypothyroidism , hyperthyroidism, hyperparathyroidism, motor variant of CIDP , infectious neuropathies like HIV and HTLV -1.

RESULTS:

Of the 49 patients, 36 (73.5%) were male and 13 (26.5%) female; 53.1% were over 50 years.

Limb weakness (85.7%)	Proximal UL (70.6%)	Proximal LL (60.8%)
	Distal UL (72.5%)	Distal LL (62.7%)
Bulbar weakness (45.8%)	Muscle wasting (56.9%)	Limb & trunk fasciculations (33.8%)
Tongue fibrillation (34.7%)	Dysarthria (42.9%)	Dysphagia (31.7%)
Shortness of breath (8.2%)	Cognitive impairment (2%)	

ALS subtypes included limb-onset (57.1%), limb-bulbar (20.4%), bulbar-onset (16.3%), pseudobulbar (6.1%), and ALS-FTD (2%). Patients with respiratory and bulbar symptoms: presented to clinic after a median duration of 12 months from onset in contrast to those without bulbar symptoms (24 months- **p-value of 0.001).**
Neurophysiological studies showed a neurogenic pattern in EMG in 100% patients. MRI brain was normal in 85.7%, with ischemic changes in 10.2%, and frontotemporal atrophy in 2%.

Revised El Escorial criteria and GOLD COAST CRITERIA both have diagnosed ALS. Revised El Escorial criteria spreads patients across multiple diagnostic certainty levels, only ~20% were “definite ALS.” Gold Coast criteria is less restrictive: all 49 patients (100%) were classified as ALS in our study. There were 4 deaths in our cohort suggestive of 2.80 deaths per 100 person-years

Criteria	Diagnosed cases of ALS	Broader Categories (Probable +Possible)	Total
Revised El Escorial	10 (20.4%) (Definitive)	39 (79.6%)	49 (100%)
Gold Coast	-	-	49 (100%)

ALSFRS- R-SE score was calculated at onset and during the last follow-up. Mean duration of follow-up: 18 months (SD 9.7). The rate of change of score was calculated. The average change for this group of patients is -0.67points/month. This indicates that, on average, the patients in this dataset experienced a decline of approximately 0.67 points in their ALSFRS- R-SE score for each month of the follow-up period.

MEAN FUNCTIONAL SCORE AT ONSET	40.5
MEAN FUNCTIONAL SCORE AT FOLLOW UP	31.3
MEAN CHANGE	9.2
MEAN DURATION	18.16 MONTHS
MEAN RATE OF CHANGE	0.67/MONTH
P VALUE	<0.001

DISCUSSION:

This study's demographic findings align well with national and international data. The male predominance (73.5% male) is consistent with the global male-to-female ratio of 2:1 to 3:1 [1, 2]. Similarly, the most common subtype, **limb-onset ALS** (57.1%), corroborates findings from large-scale studies in both India and other countries [3, 4]. **Bulbar involvement** (45.8%) as an initial symptom brings patients to the clinic earlier (**median duration 12 months compared to non-bulbar p-value of 0.0015) ,** which reinforces its significant role[5]. An observation from our data is the rate of functional decline of **0.67 points per month** on the ALSFRS-R-SE scale, which is comparable to **0.7 points per month** often cited in larger international cohorts [6]. There were 4 deaths in our cohort, suggestive of **2.80 deaths per 100 person-years** compared to **1.70 to 3.4 per 100 person-years** in other studies [3]. This finding supports the hypothesis that the disease may progress comparatively **less rapidly** in Indian patients, a theory backed by other studies that have observed a younger age of onset and potentially longer survival times in this population [1, 3]. This study's neurophysiological results, showing a neurogenic pattern in 100% of patients, are a classic hallmark of ALS and align with its core pathophysiology. However, the reported low prevalence of cognitive deficits (2%) stands in stark contrast to international research. A significant portion of ALS patients experience cognitive and behavioral changes, with up to half showing some form of frontotemporal dysfunction [5]. This highlights the need for routine/ specialized cognitive screening adapted for the Indian population.

CONCLUSION:

This study of the Indian ALS cohort confirms classic demographics (73.5% male) and neurophysiological hallmarks. Limb-onset is the most common subtype (57.1%), and bulbar involvement promotes earlier clinical presentation. Notably, the observed slower functional decline (0.67 points/month) and death rate (2.80/100 person-years) support the hypothesis of comparatively less rapid disease progression in Indian patients. The Gold Coast criteria classified 100% of patients as ALS, demonstrating its utility over the Revised El Escorial criteria enabling earlier diagnosis.

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