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DIAGNOSTIC UTILITY OF SPLIT HAND INDEX IN ALS

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ABSTRACT **Introduction:** Amyotrophic Lateral Sclerosis is a rare motor neurodegenerative disease in adults characterized by both UMN and LMN signs. Assessment of Split Hand Index through a simple electrophysiological study helps in early diagnosis of ALS. **Materials &Methods:** This is study is prospective observational study conducted in department of Neurology in tertiary referral center on patients attending OPD/In patients with clinically suspected ALS with Split Hand sign from January 2021 to August 2022. **Results:** Among 22 patients included in the study ,majority were in age group of above 66 years .In our study of 22 patients,77% of patients were males. Limb onset ALS was found in 91.9% ,bulbar onset ALS 8.1 %.In our study Split Hand Index (SHI)showed sensitivity of 75% and specificity of 100%. **Conclusions:** SHI is simple,easy,non invasive ,electrophysiological tool for early diagnosis of ALS.

KEYWORDS : Split hand sign ,Split Hand Index, Amyotrophic lateral sclerosis

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a relentlessly progressive neurodegenerative disease characterized by both UMN & LMN signs(1) There is weakness, wasting, muscle twitching of one or more limbs. Bulbar muscles may also be similarly affected.

Preferential wasting of distal hand muscles, especially thenar group of muscles, when compared to hypothenar muscles results in split hand sign clinically. The split hand sign is a specific feature of ALS(2)

This may be electro physiologically demonstrated by obtaining **split hand index** through nerve conduction study of thenar muscles: Abductor Pollicis Brevis **(APB)**, first dorsal interossei (**FDI**) & hypothenar muscles of the hand: Abductor Digiti Minimi (**ADM**) .(4)

Compound Muscle Action Potential (CMAP) of each of these muscles is obtained through stimulating these muscles through nerve conduction study.

Split Hand Index (SHI) =

$$\frac{\text{CMAP amplitude of APB muscle X CMAP amplitude of FD1}}{\text{CMAP amplitude over ADM muscle}}$$

The utility of a SHI was confirmed in a large study, with a cut-off value <5.2 with specificity 80% and sensitivity 74% in ALS when compared to ALS mimic disorder(1)

ALS is a progressive neurodegenerative disorder affecting the upper & lower motor neurons. The ALS diagnosis is delayed especially in its early stages. Preferential wasting of the thenar group of muscles, split hand sign, appears to be a specific feature of ALS. This study was undertaken to analyze the utility of split hand index in the diagnosis of ALS. It is an early, simple and non-invasive biomarker for the diagnosis of ALS.(1)

Unfortunately, no treatment has been found to halt the progression of ALS.

So, it is very important to differentiate it from other diseases which mimic ALS, like Hirayama disease, Cervical spondylosis which are potentially treatable.

MATERIALS AND METHODS

It was a prospective observational study conducted in patients with neuromuscular symptoms like wasting, weakness, muscle twitching, flailness of limbs, difficulty in swallowing who are attending a tertiary care hospital OPD or admitted in Inpatient ward of Neurology Department from Jan 2021 to Aug 2022. All the study participants were evaluated after taking informed consent ,by detailed history and clinical examination and they underwent Nerve Conduction Study of median nerve and ulnar nerve to assess the split Hand index ,MRI Brain and spinal cord were done.

Split hand index – was derived by dividing the product of CMAP amplitude recorded over Abductor pollicis Brevis muscle and first dorsal interosseius muscle by the CMAP amplitude recorded over the Abductor digiti minimi muscle.

Split hand index(SHI) =
(APB CMAP x FDI CMAP) ÷ADM CMAP

Inclusion Criteria

Patients >18 yrs of age

Patients with clinically suspected ALS according to **El Escorial criteria** with split hand sign were selected.

El-Escorial criteria(3)

• Definite ALS

Out of 4 body regions,Cranio-bulbar, cervical, Thoraccic, Lumbosacral, 3 of the body regions show both UMN & LMN signs.

• Probable ALS

Both UMN & LMN signs seen in at least 2 body regions, with UMN signs rostral to LMN signs.

• Possible ALS

UMN & LMN signs in one region

Exclusion of ALS mimics

Exclusion Criteria

Patients with history of neck trauma

Patients with sensory symptoms

RESULTS

A total of 22 patients were enrolled in the study. The results of the analysis of data collected from these participants are presented in this section.

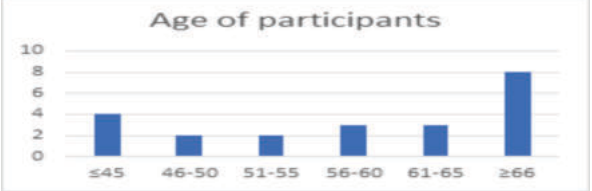

Figure 1- Distribution Of Study Participants According To Age

The age of the participants ranged from 40 to 72 years with a mean (standard deviation) age of 58.50 (10.774) years. The majority of the study participants are more than 66 years of age. The distribution of participants by age is shown in Table 1and Figure 1.

The majority of the participants were males (77.3%). Amyotrophic lateral sclerosis (ALS) is present in 45.5% of participants. The distribution of participants by ALS is shown in Figure 2.

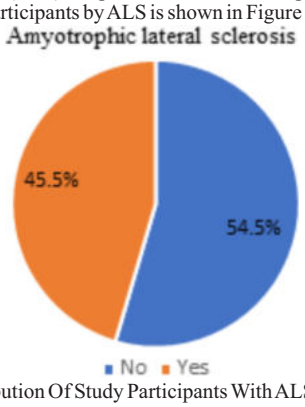

Figure 2- Distribution Of Study Participants With ALS

Of the 22 ALS patients, bulbar onset disease was observed in 2 patients (9.1%) and 36.4% have ALS present in the limbs. 18.2% have definite ALS.

Table 2- Association Of SHI With Definite ALS

Definite ALS	ALS		Total	P value
	Yes	No		
Yes	4 (100%)	0 (0.0%)	4	0.015
No	6 (33.3%)	12 (66.7%)	18	
Total	10 (45.5%)	12 (54.5%)	22	

Association of SHI with definite ALS using the Chi- square test with p value 0.015.

Table 3- Diagnostic Performance Of The SHI In ALS Patients

Parameter	Cutt-off value	Sensitivity (%)	1-Specificity (%)	AUC (95%CI)	P value
All patients (n =22) SHI	2.150 2.350	0.750 1.000	0.278 0.278	0.785 (0.597-0.973)	0.081

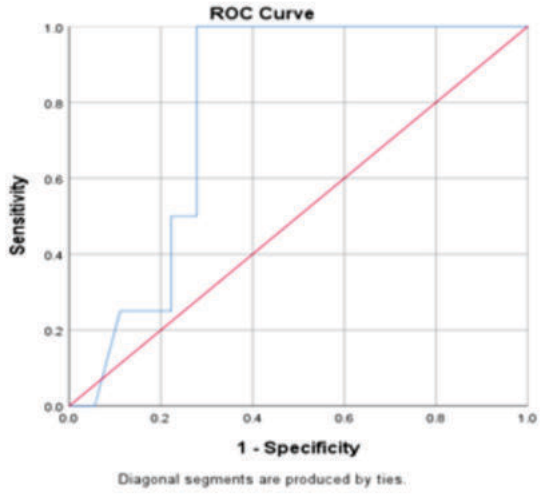

Figure 3. Receiver-operating Characteristic Curves For The SHI.

DISCUSSION

The ALS is a rare disease,with incidence and prevalence rates for sporadic ALS are almost uniform through out the world. The estimated incidence in North America & Europe is about 2 per 100,000 & prevalence is about 5-7 per 100,000 (Mehta et al). The varying symptoms and presentations make it a diagnostic challenge especially in the early stages.(6) The diagnosis of ALS still relies mainly on clinical features. The clinical criteria used in our study was Revised El Escorial Criteria.

In our study, majority of the study population ranged from 40 to 72 years of age, with a mean age of 58.50years (standard deviation of 10.774 years). The majority of participants were aged 66 years or above which is in accordance to other studies.(Mcguire & Nelson, 2006).

In our study,majority of the study population were males(77%).

In an Eastern Indian study ,M:F ratio of 4.3 : 1. (7)

In our study, mean duration of symptoms was one and a half years. In an indian study by Nalini et al, the mean duration was 17.7- 20.7 months. Another study by Norris et al revealed , mean duration of symptoms as 17 months(7) In our study, limb onset ALS cases were 8 (91.9%) and bulbar onset cases were 2 patients (9.1%). Our study had significantly higher number of Limb onset cases compared to other studies.An eastern indian study on ALS had had 43.7 % of limb onset disease and 33.3% of bulbar onset cases.

Amyotrophic lateral sclerosis (ALS) was present in 45.5% of the study participants.This indicates a substantial proportion of individuals in the study had ALS.

Kuwabara et al defined split hand syndrome as an APB/ADB CMAP ratio<0.6 or FDI/ADM ratio <0.9. Their results revealed that split hand sign was detected with 52 % sensitivity 95%specificity(14).

Menon et al, in a major study, determined that SHI could robustly differentiate ALS from mimic disorders with a sensitivity (74%) and specificity(80%).

The association analysis between SHI and definite ALS using the Chi-square test showed a statistically significant result with a p-value of 0.015. This suggests a meaningful relationship between SHI and the presence of definite ALS, which may have clinical implications.

The diagnostic performance of the SHI in ALS patients was evaluated in our study.. At a cutoff value between2.150 and 2.350, the SHI demonstrated a sensitivity of 75% and specificity of 100%.

The area under the curve (AUC) of the receiver-operating characteristic (ROC) curve was 0.785 (with a 95% confidence interval of 0.597-0.973), suggesting that the SHI has potential as a diagnostic tool for ALS patients. However, the p-value associated with this analysis is 0.081 ,which is not statistically significant at the conventional significance level of 0.05.

One possibility regarding split hand syndrome in ALS is that more frequent use of thenar muscles (APB and FDI) renders these muscles and their motor neurons more susceptible to oxidative stress and increased metabolic demand, leaving the motor neurons of these muscles more vulnerable to degeneration processes.(10)

Another hypothesis states that cortico -neuronal hyper excitability causes thenar spinal anterior horn cells to degenerate through anterograde glutamate induced excitotoxicity.

The unique pattern of muscle weakness in ALS has also been attributed to greater cortical representation of thumb and thereby stronger corticomotoneuronal connections of the affected muscle groups.(11)

CONCLUSIONS

This study provides valuable insights into the the prevalence and subtypes of ALS, and the potential diagnostic utility of the SHI. The significant association between SHI and definite ALS indicates the importance of early and accurate diagnosis. The SHI is a simple ,easy non-invasive neurophysiological test which can be used in the early diagnosis of ALS.

Further studies with a larger sample size may be needed to establish its statistical significance. These findings contribute to our understanding of ALS and may influence future research and clinical practice.

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