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"Demographic Pattern of Amyotrophic Lateral Sclerosis in Bangladesh Among Patient Admitted in a Tertiary Level Hospital"

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Abstract
Background: *Amyotrophic lateral sclerosis (ALS) is a motor neuron disease (MND) that affects both upper and lower motor neurons. The prevalence of people living with ALS, varies among geographic regions worldwide. For the betterment of the patients with amyotrophic lateral sclerosis it is very important to have the proper knowledge on the demographic pattern of amyotrophic lateral sclerosis.* ***Aim of the Study:*** *The aim of this study was to evaluate the demographic pattern of amyotrophic lateral sclerosis in Bangladesh.* ***Methodology & Materials:*** *This hospital-based prospective cohort study was conducted in the Department of Neurology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh during the period from January 2010 to December 2011. Among 56 MND patients from first screening in total 34 patients with ALS, completed the full tenure of the study were finalized as the study population. The demographic profile, clinical characteristics and risk factors were systematically recorded.* ***Results:*** *In this study among total 34 participants, the highest 41% (n=14) participants were from 41-50 years' age group. Male were 59% (n=20) and female were 41% (n=14). In total 47%, 41% and 12% participants were from lower, middle and upper class families respectively. We observed 58.82% participants had classical ALS. On the other hand, 17.65% participants had juvenile form of ALS and 23.53% participants had Monomelicamyotrophy. Cognitive impairment was found negative among all the participants. Both UMN and MLN were found among 100% patients.* ***Conclusion:*** *In this study we found gender and age of the participants was potential aspect regarding amyotrophic lateral sclerosis (ALS) disease. All the findings of this study may be helpful for farther similar studies and in the treatment procedure of ALS.*
Key words:*ALS, sclerosis, amyotrophic.*

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I. Introduction

Amyotrophic lateral sclerosis (ALS) is a type of motor neuron disease (MND). Motor neuron disease (MND) is used to designate a progressive degenerative disorder of motor neurons in spinal cord, brainstem and motor cortex manifest clinically by muscular weakness, atrophy and corticospinal tract signs in varying combinations. It is a disease of middle life, for the most part and progress to death in a matter of 2 to 6 years or longer in exceptional cases. MND's are progressive diseases of adults resulting from variable degeneration of the upper motor and lower motor neurons. The recognized types, which display considerable clinical similarity, are sporadic,[1] familial and western pacific forms.[2] The diagnostic criteria of the disease have been established by the World Federation of Neurology Subcommittee on motor neuron diseases. Its clinical variants result either from predominant degeneration of the LMNs of limb muscles (PMA) or bulbar muscles (progressive bulbar palsy, PBP) or the UMNs (primary lateral sclerosis and progressive pseudobulbar palsy). However, these forms may be distinguishable only early in the disease, as most patients eventually develop clinical and pathological features of UMN and LMN involvement. This is a common disease, with an annual incidence rate of 0.4% to 1.76% per 100,000 populations. Men are affected somewhat more frequently than women. Male to female ration is approximately 1.6:1. Most patients are more than 50 years old at the onset of symptoms and the incidence increase with each decade of life. The disease occurs in a random pattern throughout the world except for a

dramatic clustering for patients among inhabitants of the Kiipeninsula in Japan and Guam, where MND is often combined with dementia and Parkinsonism.[3] MND is characterized by the gradual death of upper and lower motor neurons leads to loss of motor function and clinical syndrome of muscle weakness, wasting, and paralysis resulting in death typically within 2–3 years.[4] Generally, the presentations include ALS, in which both the lower motor neurons and upper motor neurons are affected. It is frequently referred to as "Lou Gehrig's disease" in memory of the famous baseball player who died of ALS[5] in 1941. Other forms primary lateral sclerosis, which is less common than other presentation with only upper motor neuron involvement; and progressive muscular atrophy, with only lower motor neuron signs. Population-based studies have established that the incidence of ALS in Europe is fairly uniform at 2.16%/100,000 person-years.[6]Although ALS affects people worldwide, an exact incidence of this disease is not yet known. Men have a higher incidence of disease (3.0%/100,000 person-years; 95% confidence interval [CI] 2.8%–3.3%) than do women (2.4%/100,000 person-years; 95% CI 2.2%–2.6%). Besides these, genetic, environmental, occupational, toxins, high-level physical activity, electrical injury, physical trauma, medical illness, and high magnetic field exposed activity; all have been suggested by different investigators as causal factors at different times but not proved yet with certainty.[7,8] There are very few information regarding demographic pattern of the disease. Riluzole, a glutamate antagonist, is the only drug approved by the Food and Drug Administration for the treatment of ALS. In two therapeutic trials, riluzole prolonged survival by 3–6 months.[9,10]One of the biggest challenges today is the well-established heterogeneity of ALS,[11] with patients displaying widely different patterns of disease manifestation and progression, and genetic analyses suggesting heterogeneity of the underlying biological mechanisms.[12] This heterogeneity has detrimental effects on clinical trial planning and interpretation,[13] on attempts to understand disease mechanisms, and on clinical care, as it increases uncertainty about prognosis and optimal treatment. Thus, successfully stratifying ALS patients into clinically meaningful sub-groups can be of great value for advancing the development of effective treatments and achieving better care for ALS patients.

II. Methodology & Materials

This hospital-based prospective cohort study was conducted in the department of neurology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh during the period from January 2010 to December 2011. Among 56 motor neuron disease (MND) patients from first screening in total 34 patients with ALS, completed the full tenure of the study were finalized as the study population. The demographic profile, clinical characteristics and risk factors were systematically recorded.The intervention had been approved by the ethical committee of the mentioned university. The demographic profile, clinical characteristics and risk factors were systematically recorded and these patients were followed for next 1 year. The data related to sociodemographic profile, medical history, risk factors, neurological examination parameters and treatment practices were collected systematically at the time of enrolment in the study using predesigned structured data recording format. The electromyography (EMG), nerve conduction study (NCS) and magnetic resonance imaging (MRI) were done for every patient.Data regarding age of onset, progression of symptoms, family history of disease and initial symptoms: the presence of fasciculation's, dysphagia, dysarthria, cramps, muscular weakness, emotional lability and localization of disease onset (bulbar vs. limb onset ALS) were collected.Besides these, ALS risk factor questionnaire was obtained by personal interview to obtain self-reported lifetime residential and occupational history and personal lifestyle factors. Risk factors studied were as follows (a) History of trauma requiring admission to hospital, (b) Family history of MND, (c) History of electric shock, (d) History of smoking cigarettes, (e) History of living >12 months in a rural area, (f) History of farm work for >12 months, (g) History of possible occupational or hobby exposure to lead. (Construction batteries, automobile industries, paints, as soldering materials in electronics), (h) History of pesticide or herbicide use.All cases were classified according to El Escoreal criteria into definite, probable and possible ALS. ALS functional rating scale (ALSFRS) score calculated at the time of presentation and after 1 year of follow up.The descriptive statistics of the study population were reported as counts and percentages for categorical variables and mean ±SD for continuous variables with normal distribution.

III. Results

In this study among 56 MND patients from first screening in total 34 patients with ALS, completed the full tenure of the study were finalized as the study population. In analyzing the age of the participants, the highest 41% (n=14) participants were from 41-50 years' age group followed by 20%(n=7) from >70 years' age group, 15%(n=5) were from 61-70 years' age group, 12%(n=4) from 51-60 years' age group 9%(n=3) were from 31-40 years' age group and the rest 3%(n=1) were from <30 years' age group.According to the patient's gender we found that, most of the patients were male which was 59%(n=20). On the other hand, only 41%(n=14) of the total participants were female. So male was dominating in number here.In analyzing the occupational status of the participants, we found the participants belonged several professions. Those were farmer 18%(n=6), day laborer 18%(n=6), student 15%(n=5), general service 12%(n=4), defense service

9%(n=3) businessman 9%(n=3), teacher 6%(n=2), housewives 6%(n=2), athletes 6%(n=2) and finally lawyer 3%(n=1). When we collected the family history we found 100% participants were free from any family history of ALS. In this study, the highest number of patient were from lower class family which was 47% (n=16). Then 41.18% were from middle class and only 11.76% were from upper class families.In analyzing the types of ALS of the participants we found, 58.82% participants had classical ALS whose mean age was 53±7.12 years and here the male-female ratio was 1.44:1. On the other hand, 17.65% participants had juvenile form of ALS whose mean age was 21±4.12 years and the male-female ratio was 8:1. Besides these, 23.53% participants had Monomelicamyotrophy (MMA) whose age mean was 24±8.6 years and here the male-female ratio was 3:1. Cognitive impairment were found negative among all the participants. Both upper motor neuron (UMN) and lower motor neuron (LMN) were found among 100% patients.

Table-1 Patients Age WiseDistribution (N=34)

Patients Age Group (years)	Frequency (n)	Percentage (%)	Mean Age
<30 years	1	3.0	53±7.12 years
31-40 years	3	9.0	
41-50 years	14	41.0	
51-60 years	4	12.0	
61-70 years	5	15.0	
>70 years	7	20.0	
Base	34	100.0	

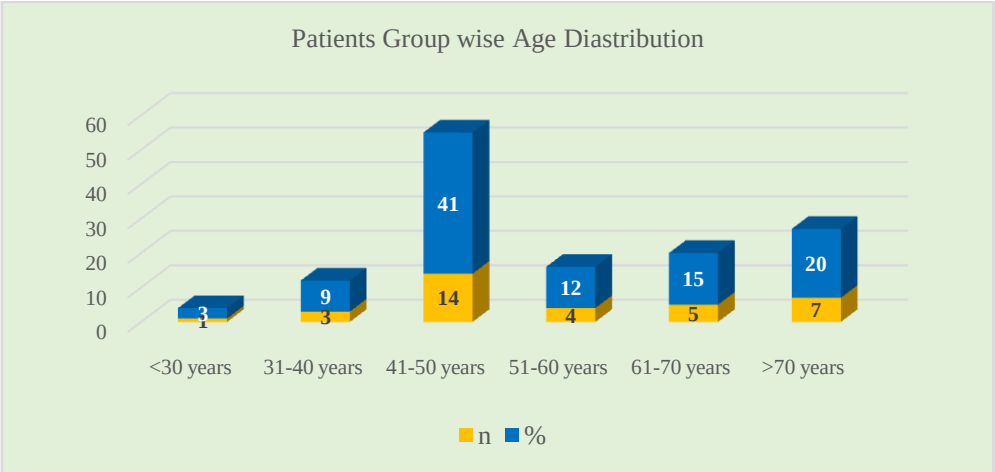

***Figure: I** Patients Age Distribution (N=34)*

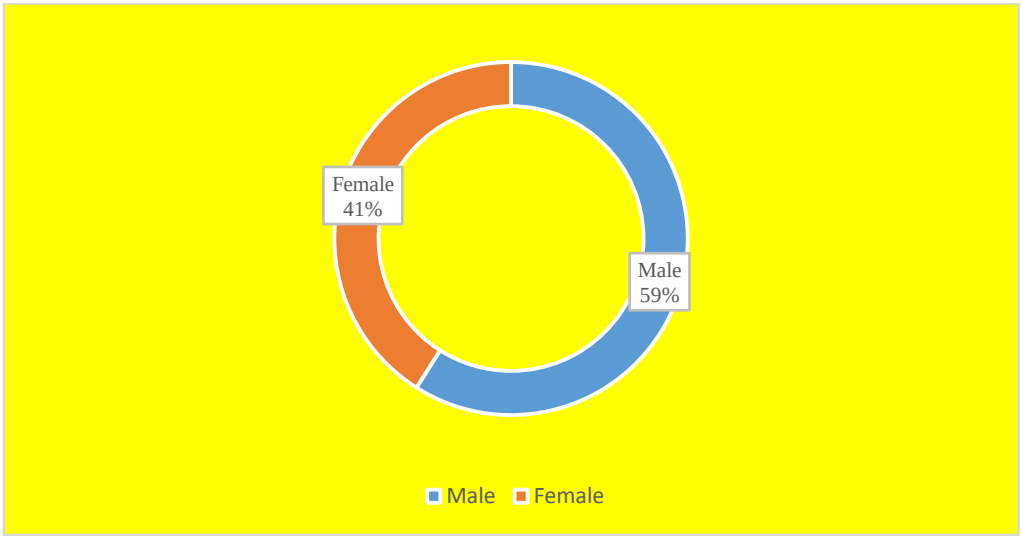

***Figure: II** Gender Wise Patients Distribution (N=34)*

Table-2 Patients Age Wise Distribution (N=34)

Patients Occupations	Frequency (n)	Percentage (%)
Farmer	6	18.0
Day Laborer	6	18.0
Student	5	15.0
General service	4	12.0
Defense service	3	9.0
Businessman	3	9.0
Teacher	2	6.0
Housewives	2	6.0
Athletes	2	6.0
Lawyers	1	3.0
Base	34	100.0

Figure: III *Patients Occupation Wise Distribution (N=34)*

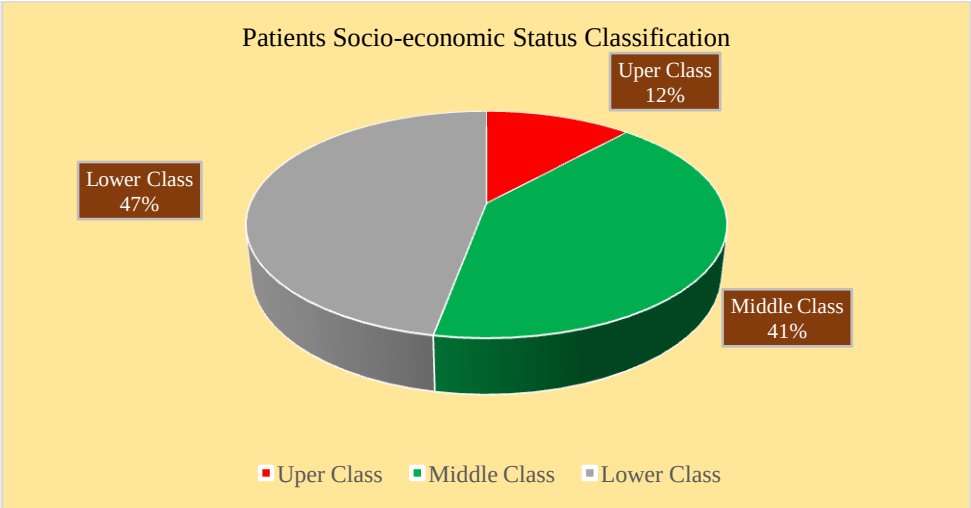

Figure: IV *Patients Socio-economic Status (N=34)*

Table-3 ALS types Among Participants (N=34)

Patients Age Group (years)	Frequency (n)	Percentage (%)	Mean Age	M:F
Classical ALS	20	58.82	53±7.12	1.5:1
MMA	8	41.0	21±4.12	8:1
Juvenile ALS	6	17.65	24±8.6	3:

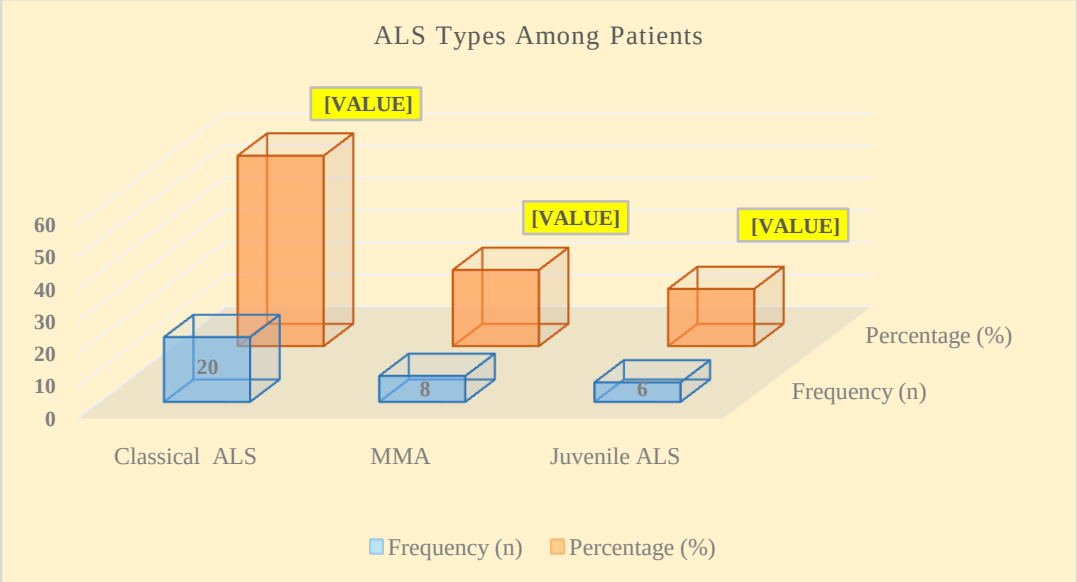

***Figure: V** ALS types of patients (N=34)*

IV. Discussion

This hospital-based prospective cohort study was conducted in the Department of Neurology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh during the period from January 2010 to December 2011. The intervention had been approved by the ethical committee of the mentioned university. Among 56 Motor neurone disease (MND) patients from first screening in total 34 patients with ALS, completed the full tenure of the study were finalized as the study population. The demographic profile, clinical characteristics, and risk factors were systematically recorded.The purpose of this study was to evaluate the demographic pattern of amyotrophic lateral sclerosis in Bangladesh.In this study among 56 MND patients from first screening in total 34 patients with ALS, completed the full tenure of the study were finalized as the study population. In analyzing the age of the participants, the highest 41%(n=14) participants were from 41-50 years' age group followed by 20%(n=7) from >70 years' age group, 15%(n=5) were from 61-70 years' age group, 12%(n=4) from 51-60 years' age group 9%(n=3) were from 31-40 years' age group and the rest 3%(n=1) were from <30 years' age group. According to the patient's gender we found that, most of the patients were male which was 59%(n=20). On the other hand, only 41%(n=14) of the total participants were female. So male was dominating in number here. So the male-female ratio was 1.44:1. A study from Northern India with 62 cases of ALS showed a M: F ratio of 4.6:1, mean age of onset of 46.9 years, limb onset in 70.9%, bulbar onset in the remaining.[14]These findings have huge dissimilarities with ours'. We found suchdissimilarities in another study, Rai and Jolly reported a M: F ratio of 6:1 among their 70 cases with ALS which was significantly high as compared to our series.[15]But in another study we found similarity in male female ratio. In yet another study from West India 82 cases of ALS had aM: F ratio of 2:1 and a high proportion had onset before the age of 40 years.[16]In analyzing the occupational status of the participants, we found the participants belonged several professions. Those were farmer 18%(n=6), day laborer 18%(n=6), student 15%(n=, service holder (12%), businessman (9%), defense person (9%), teacher (6%), housewife (6%), athlete (6%) and lawyer (3%). A significant number of the ALS patients in the study were the rural domicile people and were farmers. It has been detected as a risk factor of ALS in several studies.[17]Rural people are exposed to several chemicals used in the agricultural fields land may have repeated physical trauma arising from their physical nature of jobs.In this study, the highest number of patient were from lower class family which was 47%(n=16). Then 41.18% were from middle class and only 11.76% were from upper class families. According to the findings we think socioeconomic class, living slandered and gender are most potential factors for ALS. Females were having low ALSFRS score as compared to males, more number of deaths and early mean age at the time of death were found in several international studies. From these parameters, it can be concluded that females have worse prognosis than males. Results are similar to study[18] "Prognosis in ALS" done in Washington in 180 patients of ALS, which found that, older age and female sex were strongly associated with poor survival.

V. Conclusion

In this study we found gender and age of the participants was potential aspect regarding amyotrophic lateral sclerosis (ALS) disease. All the findings of this study may be helpful for farther similar studies and in the treatment procedure of ALS.

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