Public Awareness of Motor Neuron Disease: A Cross-Sectional Study

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Submission: October 20, 2020; **Published**: November 18, 2020

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Abstract

Background: Motor Neuron Disease (MND) is an assortment of clinically analogous ailments that instigate progressive degeneration of motor neurons in the central nervous system. This study aims to evaluate the awareness and knowledge of MND in the general population of Karachi, Pakistan.

Methods: 680 individuals were initially approached for data collection, out of which 633 fit the inclusion criteria (response rate= 93.01%). Nonprobability convenience sampling was utilized to select participants who fell under the age group of 15-75 years, were residents of Karachi and had no prior formal education in the fields of medicine, dentistry, pharmacology or nursing. Simple statistical analysis for frequency was put into play, with Chi-Square test to gain association between the variables.

Results: Out of 633 questioned participants, 235 subjects acknowledged being familiar with MND; majority fell in the moderate awareness category (34.46%). Nearly one-third of the individuals from High awareness category identified Amyotrophic Lateral Sclerosis (ALS) as the most common MND. The analysis showed a significance with age (p=0.043), gender (p=0.001), source of knowledge (social media and publications, p=0.001), cognition regarding Stephen Hawking's battle with ALS (p=0.001), the ice bucket challenge (p=0.001) and having family or friends suffering from MND (p=0.001).

Conclusion: In conclusion, only one third of those included had awareness of MND. A surprising majority of unaware participants claimed recognition of Stephen Hawking, his ailment and the ALS ice bucket challenge. This further creates the need for public awareness of the said disease.

Keywords: Motor neuron disease; MND; Amyotrophic lateral sclerosis; ALS; Lou Gehrig's disease; Stephen Hawking; Ice Bucket Challenge

Abbreviations: MND: Motor Neuron Diseases; ALS: Amyotrophic Lateral Sclerosis

Introduction

Motor Neuron Disease (MND) is an assortment of clinically analogous ailments that instigate progressive degeneration of motor neurons in the brain and spinal cord. In conjunction with male preponderance, the majority of individuals hail from fifth to seventh decade of life at the time of diagnosis [1]. Recently it has been brought to attention that the total cost per patient with MND exceeds that of any other neurologic condition, for example; Parkinson's disease, dementia or stroke [2]. The reported incidence rates of MND have shown vast discrepancies on account of the diverse methodological approaches employed [3]. However, population-based studies conducted in Europe repeatedly provided an incidence rate of 2 per 100,000 people per

annum and yet there are numerous disputes concerning explicit epidemiology of MND [4,5]. The Global Burden of Disease (GDB) report about MND in Pakistan stated a death count of 233 and the Disability-Adjusted Life Years (DALYs) were calculated to be 8950 in 2016 alone [6]. MND is classified into four major types, based on the variety of neurons compromised along with its inception in whichever part of the body; Amyotrophic Lateral Sclerosis (ALS), Progressive Bulbar Palsy (PBP), Progressive Muscular Atrophy (PMA) and Primary Lateral Sclerosis (PLS). The most common MND is ALS, also known as Lou Gehrig's disease and was named after a famous baseball player. ALS endangers both upper and lower motor neurons with predominant weakness and wasting of muscles of the limbs. The reckoned lifetime risk of ALS is one in

400 [7]. ALS is a physically debilitating disease and a public health concern; it significantly affects the economy and in 2015 alone, the estimated burden was found to be US$ 279-472 million in United States [2]. The life expectancy of a patient afflicted with ALS is supposedly three to five years after diagnosis [8]. However, the world witnessed the unrivaled subsistence of renowned physicist Stephen Hawking for a spell of over five decenniums after his diagnosis was concluded. No comprehensible reasoning has been proposed to elucidate this anomalous turn of events. Currently, no specific tests exist for the conclusive diagnosis of MND and the pursuit for a biomarker is still underway; hence, this entity is diagnosed following exclusion of other neurological diseases [9]. Unfortunately, the sole drug to show promising results- Riluzole- prolongs the life expectancy for only three to four months and notifying the patients and his/her family members of the incurable nature of MND remains the most distressing task [10]. In addition to physical impairment, MND psychosocially disables the patient and the trivial to absent levels of awareness of the disease causes patients and their families to struggle with this revelation [11]. Scarcity of pertinent research further impedes the appreciation of MND as a public health issue requiring immediate attention. Consequently, public awareness is an imperative tool to aid MND patients and their families to withstand this calamity and make au courant decisions regarding their condition. Thus, we conducted this study with the primary objective to evaluate the awareness of MND in the general population of Karachi. Our secondary objective was to assess the knowledge in the aware group and find its association with the demographics, familiarity with Stephen Hawking, ALS ice bucket challenge and the respective sources of knowledge.

Materials and Methods

Study design and sample

A study that commenced in February 2018 for a period of 15 months, ending in May 2019, was based principally to evaluate the awareness of MND among the general population of Karachi, Pakistan. It was conducted by a group of medical students of Dow University of Health Sciences (DUHS) after having sought sanction from the respective Institutional Review Board. The sample size was calculated to be 600 through Open Epi version 3.01 using a confidence level and confidence limit of 97% and 5% respectively. Nonprobability convenience sampling was utilized to select participants who fell under the age group of 15-75 years, were residents of Karachi, Pakistan and had no prior formal education in the fields of medicine, dentistry, pharmacology or physiotherapy, so as to prevent overestimation of knowledge amongst the general population. There was no discrimination based on gender, ethnicity, socioeconomic status or religion. Awareness, knowledge and attitudes towards the disease were assessed, with awareness referring to acknowledgement of the existence of the disease or problem, knowledge being the accuracy of the target's information or facts regarding the disease and attitudes alluding to the perceptions of the people pertaining to the matter at hand

[12].

Data collection and Administration

A standard, self-administered questionnaire was distributed amongst the participants after gaining their written consents, in the languages commonly spoken in Pakistan in order to remove the interviewer and procedural bias. Maximum precautions were practiced so as to remove any kind of selection or response bias. Among the 23 questions asked in each interview, most were close-ended and comprised of dichotomous, trichotomous or multiple options. Out of the 680 questionnaires distributed, 17 were excluded either due to incomplete information or not conforming to the inclusion criteria, and 30 peoples refused to fill the in the responses; thereby, yielding a response rate of 93.01%. A major part of the questionnaire was composed of questions to determine the acquaintance with disease etiology, symptoms, associated risk factors, gender prevalence, management and prognosis from those who claimed to be aware of MND. The questionnaire also assessed the participants' knowledge of awareness from public events such as the ALS ice bucket challenge and if they had any prior knowledge regarding Stephen Hawking's plight.

Knowledge assessment

14 queries from the questionnaire aimed to assess the knowledge of the interviewee. A score of 1 was allotted to each correct answer while each incorrect answer scored a 0, amounting to a minimum and maximum possible score of 0 and 14 respectively. Based on the 25th and 75th percentile of the cumulative score, three ranges were obtained low (0-2), moderate (3-5) and high (6-14).

Data Analysis

Statistical Package for Social Sciences (SPSS) version 24.0 (IBM Corp. Armonk New York) was employed to enter and analyze the data. Chi-Square test was applied to assess the categorical data and a p-value of less than 0.05 was considered significant. Median values with Interquartile ranges or mean values with standard deviations were used to process numerical data keeping in consideration the asymmetry of the distribution.

Results

Characteristics of study population

The sociodemographic facets of the participants are demonstrated in table 1. Concisely, majority of the interviewees were females (n=388, 61.3%) and hailed from 15 to 30 years of age (n=525, 82.93%). Mean age of the participants was 25.42 ± 9.49 (ranging between 15-75). Approximately, 541 participants (85.5%) were high school graduates.

Knowledge prevalence and questions addressed

In order to evaluate the awareness regarding MND, the targeted population was asked if they were familiar with the term MND which resulted in a cognizance rate of only 37.12% (n=235).

A detailed questionnaire was formulated to extract the extent of knowledge in these 'aware' participants based on the number of correct answer given for each query. According to the scores attained, participants were divided into three categories; high, moderate and low. Even though 235 subjects acknowledged being familiar with MND, a major bulk of them did not have sufficient knowledge regarding it hence falling in the moderate category (n= 81, 34.46%) followed by low (n= 78, 33.19%) and high (n= 76, 32.34%) categories. Participants' perception regarding the fundamental aspects of MND was analyzed as illustrated in table 2, with the underlined options exhibiting the correct answer. As shown in table 2, nearly one third of the individuals from the high category recognized ALS as the most common MND whereas a vast majority from not only the high awareness category but also from the moderate and low groups left it unanswered.

Table 1: Association of knowledge with sociodemographic characteristics and awareness regarding Stephen Hawking and ALS ice bucket challenge.

Parameters	High knowledge	Moderate knowledge	Low knowledge	No knowledge	P-value[a]
Age ranges					0.043
15-30	64	70	73	318	
31-45	8	9	3	47	
46-60	4	1	2	29	
61-75	-	1	-	4	
Gender					0.001
Female	62	58	54	214	
Male	14	23	24	184	
High school graduate					0.46
Yes	67	71	66	337	
No	9	10	12	61	
Was social media your source of knowledge?	29	44	31	-	0.001
Were publications your source of knowledge?	31	30	32	-	0.001
Do you know Stephen Hawking?					0.001
Yes	70	60	47	113	
No	6	21	31	285	
Death of Stephen Hawking[b]					0.088
Before	19	9	11	60	
After	57	72	67	338	
Heard of or participated in ALS ice bucket challenge?					0.001
Yes	37	54	36	137	
No	39	27	42	261	
Had family or friends suffering or suffered from MND[c]					0.001
	21	5	4	-	

[a] A p-value of less than 0.05 is considered significant.
[b] Frequencies according to the data collected before and after Stephen Hawking's death.
[c] Motor Neuron Disease.

Table 2: Extent of knowledge regarding Motor Neuron Disease (MND)[a]

Questions asked:	High Knowledge	Moderate Knowledge	Low Knowledge
Did you know the risk to get MND is 1 in 400?			
Yes	34	19	3

No	42	62	75
What is the most common MND?			
Amyotrophic Lateral Sclerosis	21	6	1
Others	8	3	2
Unanswered	47	72	75
What causes MND?[b]			
Cause is unknown	15	3	1
Combination of multiple factors	29	25	18
Environmental factors	21	21	11
Genetic factors	32	38	28
Heavy metals	5	12	3
Lifestyle	5	13	4
Don't know	13	19	36
What is the classical symptom of MND?			
Shaking and tremors	15	11	14
Problem in balance or incoordination	31	15	7
Memory deterioration	5	6	3
Headache	1	5	7
Problem in movement	18	28	12
Difficulty in speech	4	6	3
Don't know	2	10	32
Is MND difficult to diagnose?			
Yes	46	21	8
No	21	34	14
Don't know	9	26	56
What is the life expectancy after diagnosis?			
Less than 5 years	40	13	4
5 to 10 years	16	21	6
Normal life expectancy	4	12	9
Don't know	16	35	59
Is MND a fast progressing disease?			
Yes	59	29	11
No	8	22	10
Don't know	9	30	57
Are effective treatments available for MND?			
Yes	9	17	19
No	61	43	7
Don't know	6	21	52
Who can best provide care for people with MND?b			
Physician	3	2	1
Neurologist	38	45	46
Psychiatrist	1	1	4
Multidisciplinary	31	23	5

Don't know	3	10	22
Does MND affect senses and intellect of an individual?			
Yes	41	47	40
No	29	21	6
Don't know	6	13	32
Which gender does MND affect more?			
Females	6	5	5
Males	30	18	3
Both equally	23	28	26
Don't know	17	30	44
Which age group is most likely to suffer from MND?			
30 to 50	24	21	14
50 to 70	32	30	10
Above 70	5	4	5
Don't know	15	26	49
Is MND a poorly explored disease?			
Yes	33	40	17
No	16	22	25
Don't know	27	19	36
Is MND similar to any of the following?			
Alzheimer's disease	4	17	8
Parkinson's disease	41	28	16
Neither	17	12	7
Don't know	14	24	47

[a]Underlined options represent the correct answers.

[b]Frequencies for separate options do not add up to 235 as people were given a chance to choose multiple answers.

The causative factor for MND given by majority from each of the category was genetic factors further proving insufficient knowledge in this field. Another misconception that was brought into light was the consideration of a neurologist as the sole care provider in the context of the disease in all the three awareness groups. In addition to this, MND was thought to be a disease primarily affecting the senses and intellect of the sufferers in all the groups. It was however accurately presumed by a significant number of participants to be a fast progressing and poorly explored entity that is difficult to diagnose. As demonstrated in table 1, the knowledge of MND had a statistical significance with ages 15-30, females, sources of knowledge (social media and publications), awareness regarding Stephen Hawking's battle with ALS, the ice bucket challenge and having afflicted family or friends (p-values <0.05). Nearly all the individuals with high knowledge and three-fourth with moderate level of knowledge were specifically found to be more familiar with Stephen Hawking and his long term illness (p value= 0.001). Surprisingly, 38.93% and 51.1% people were reported to be familiar with Stephen Hawking and ALS ice bucket challenge, respectively despite being unaware of MND.

Discussion

Uncommon, uncharted and unrelenting, such is the curious case of MND - a conglomerate of conditions that contributed to about 0.04% (910,000) of the global DALYs in the year 2015 [13]. This percentage, although seemingly frivolous, is inclined to increase as the years go by. According to a study conducted in 2015, the world over the course of 2.5 decades shall witness a 69% increment in the number of cases of ALS, with an upsurge of 81% in Asia alone. Furthermore, majority of the burden of ALS will be gradually translocated to developing countries, on account of their escalating ageing population [14]. These alarming statistics coupled with the overwhelming emotional, financial, physical, psychological and social burden of the disease on the sufferers, their families and the community warrants a baseline evaluation of awareness, knowledge and attitudes amongst the general masses of a developing nation. Consequently, this study was conducted, yielding an awareness of 37.1%, with 67.6% (n= 159) of those having negligible to mediocre knowledge. A similar lack of awareness and knowledge was reported in a letter to the

editor by Davies et al in 2010 [11]. This similitude of results almost a decade later can be attributed to the lower literacy rates, lack of awareness campaigns and pertinent educational and social reforms in our region. Furthermore, those struck by this disease experience a loss of their social standing, independence and sense of normality, leading to deliberate avoidance of social gatherings for fear of reliance on others, being seen in a wheelchair in public and lisping speech. Additionally, the culture of endless interrogation and unneeded recommendations in our society compel such individuals to undergo incarceration in their homes. As a consequence, the people in their circle and general surroundings are ignorant, harbor misconceptions and never truly understand the devastating implications that this disease entails. Over one half of the 'aware' participants were under the notion that those afflicted with MND were to be looked after solely by a neurologist. In accordance with the National Institute for Health and Care Excellence (NICE) guidelines, however, such patients are ministered to by a specialized multidisciplinary team, encompassing the disciples of neurology, palliative care, nutrition, nursing, physiotherapy, occupational therapy, respiratory physiology and speech-language pathology [15]. A striking magnitude of 'aware' respondents deem MND akin to Parkinson's disease and Alzheimer's disease. All three of these infirmities are hyponyms for neurodegenerative diseases, sharing core pathological attributes at a cellular and subcellular level while simultaneously differing in their clinical presentation [16]. Additionally, 51.5% of those 'aware' correctly presumed MND to be an unexplored entity. Ever since its introduction in 1874, MND remains an enigma by virtue of its etiology, pathogenesis, diagnosis, modality of treatment and life experiences of those affected and their carers [17]. Virtually all of the individuals with high level of knowledge and approximately three-fourth of those with moderate level of knowledge significantly affirmed cognizance of Stephen Hawking and his long-term ailment (p-value=0.001). This can be accredited to the unprecedented 86-fold increase in the search interest on Google in Pakistan in March 2018, as a stark majority of our data was collected following his demise [18]. Nearly two-fifth of the respondents acknowledged familiarity with the ALS ice bucket challenge, with more than 50% of them astonishingly being unaware of MND. In the summer of 2014, the ALS ice bucket challenge garnered global recognition following its launch on social media by renowned celebrities and politicians in order to raise funds for its research. Our respondents, however, in spite of needless indulgence in social networking and being acquainted with the ALS ice bucket failed to correlate it with MND- a blanket term for ALS and other similar diseases.

Recommendations

Raising awareness amongst the general population by means of educational strategies, campaigns and social programs hosted by a native spokesperson, is an intervention of paramount significance and its benefits are three-fold. Firstly, these strategies will aid the patients and their family members in making an informed decision in the face of conflicting choices regarding their treatment. Secondly, the community care workers will register the pressing need for provision of support to such patients. Lastly, the educated masses can donate to relevant associations and these proceeds will pave the way for groundbreaking research that will ultimately translate into a cure for this disease.

Limitations

The data for this study was obtained via non probability convenience sampling and since this technique is liable to selection bias, the resultant skewed sample does not depict an accurate estimation of awareness and knowledge amongst the target population. Secondly, our study failed to assess the influence of socioeconomic status and occupation on awareness and knowledge and this gap in information will be fairly challenging to transcend when developing educational programs targeting people hailing from heterogeneous social and occupational backgrounds. Lastly, the unexpected and unfortunate passing of Stephen Hawking in the midst of our data collection may have led to a cognitive bias by raising insight in those with prior knowledge and overall awareness in general, causing a further distortion of the results.

Conclusion

To conclude, this study demonstrates an unfortunate and deplorable level of awareness, lack of knowledge and ignorant attitudes in the general population regarding MND, signifying the dire need for further research on the topic and establishment of an awareness program to educate people which could aid patients and their family members to better understand the disease and cope with it, and the society to recognize the debilitating nature of the illness and offer support to the devastated souls.

Acknowledgement

Conflict of interest

The authors declare that there is no conflict of interest.

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