Validation and psychometric properties of the Turkish version of Neuromuscular disease Swallowing Status Scale (NdSSS) in patients with oro-pharygo-esophageal dysphagia in neuromuscular disorders

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Abstract

Objective: **Dysphagia is one of the most disabling conditions arising from neuromuscular disorders(NMD). There is no specific methods to use in the evaluation of dysphagia in NMD patients. We aimed both to evaluate the applicability of the Neuromuscular Disease Swallowing Status Scale (NdSSS) for dysphagia in all phases of swallowing in various NMD patients and to investigate psychometric properties of this scale.**

Methods: **Patients with NMD were enrolled. Functional Oral Intake Scale (FOIS), Fiberoptic Endoscopic Evaluation of Swallowing (FEES), NdSSS and High-Resolution Esophageal Manometry (HRM) were performed on all subjects within 72 hours. While the convergent and concurrent validities were used as validation method, Cohen's kappa and Cronbach's alpha coefficient were calculated for inter-rater reliability. The correlation between FOIS, PAS and HRM diagnosis according to Chicago version 3.0 (CCv3) were analyzed.**

Results: **115 NMD patients were included. There was good correlation between NdSSS and FOIS and PAS scores (Spearman's rank correlation coefficient (r):0.927, r:0.927 and r:-0.836, r:0.841, respectively). Also, there was a positive good correlation between NdSSS and CCv3 evaluating disorders of esophageal peristalsis (r:0.677-0.679, p=0.001). When evaluated separately, there were good correlation between NdSSS levels; and PAS (r:-0.648-0.656); and CCv3 (r:0.514-0.573) levels for ALS. For Myasthenia gravis there was a good correlation between NdSSS levels; and CCv3 (r:0.577-0.622); FOIS (r:0.508-0.521); and PAS (r:-0.504-0.519) scores. Also, for myopathy; a very good(CCv3(0.976-0.982)) and good(FOIS (0.511-0.581) and (PAS (-0.516-0.550)) correlations were defined for myopathy.**

Conclusion: **The NdSSS was found applicable to detect both oropharyngeal and esophageal dysphagia risk in patients with NMD and is a valid and reliable swallowing screening tool that can evaluate oro-pharyngo-esophageal dysphagia in NMD patients.** (Acta gastroenterol. belg., **2022,** 85, **21-27**).

Keywords: Dysphagia, neuromuscular disorders, neuromuscular disease swallowing status scale, NdSSS, myasthenia gravis, amyotrophic lateral sclerosis, non-inflammatory myopathy, high resolution manometry.

Introduction

Swallowing is a complex event that occurs with the voluntary and reflex activities of more than 30 muscles and nerves. It consists of three phases as oral, pharyngeal and esophageal swallowing phases. Dysphagia is a difficulty in swallowing caused by any disorder along the way from mouth to stomach (1).

Dysphagia is one of the most disabling conditions arising from neuromuscular disorders (NMD), and it can be seen in almost all patients according to type and the stage of the disease (2-4). Problems of the oral phase, where muscle activity is at the forefront, such as difficulty in chewing and preparing a bolus are frequently seen in these patients (5,6). The increase in mortality and morbidity is generally due to aspiration and aspiration pneumonia caused by pharyngeal phase disorder (7). While oropharyngeal dysphagia is common in some subgroups of NMD patients such as amyotrophic lateral sclerosis (ALS), inclusion body myositis and muscular dystrophy, esophageal dysphagia is more common in myasthenia gravis (MG) and non-inflammatory myopathy(2-4,6,9,10), in other words, all three phases of swallowing can be affected in NMD patients.

Early dysphagia diagnosis can reduce the risk of complications such as malnutrition, dehydratation, aspiration pneumonia, and death, and can increase the health related quality of life.

Instrumental methods such as videofluoroscopy, endoscopy, barium radiography and manometry are used as the criterion standards for the diagnosis of dysphagia (11-13).However, these methods are expensive methods requiring special equipment and trained personnel. Therefore, screening tests are recommended as the first-line methods to detect the dysphagia risk because of their easy and, repeatable, cost-effective and non-invasive characteristics (14). Bedside screening tests are recommended as the first line in the guidelines in neurological diseases such as stroke (15). Because when the complications of dysphagia are considered, the fastest and most effective intervention is life-saving. Especially in stroke, many bedside screening tests have been developed and actively used. However, there is currently no specific methods to use in the evaluation of dysphagia risk in all NMD patients. Although there is a screening test developed for the detection of dysphagia

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risk in ALS patients, it is a disease specific test and is not favorable for other NMD patients (16). Neuromuscular disease swallowing status scale (NdSSS) is a screening test developed to show dysphagia risk status in various NMD (17). On the other hand, the reliability and validity of NdSSS were only performed for Duchene muscular dystrophy (DMD) and ALS patients. Also, there is no screening test to detect of dysphagia risk in NMD patients in our country. There is need for further research to assist in the development of definitive screening tests for this population.

For these reasons, the present study aimed both to evaluate the applicability of the NdSSS for dysphagia risk in all phases of swallowing in various NMD patients and to investigate psychometric properties of this scale for our country.

Methods

Study Design

This study was performed at neurology, gastro-enterology, physical medicine and rehabilitation and otolaryngology clinics of our hospital between October 2018 and August 2020.

A total of 124 NMD patients who were admitted to neurology clinic and consulted to the gastroenterology, otolaryngology and PMR clinics were enrolled in the study. Endoscopic evaluation was performed in all patients who accepted the study to exclude mechanical problems that could cause dysphagia. Nine patients who refused or who cannot to go for manometric evaluations were excluded. A total of 115 patients were included in the study.

Patients

Patients aged between 18-75 years, who had a confirmed diagnosis of NMD by a neurology specialist, who had at least one-year duration for disease, and had no swallowing rehabilitation in the last 6 months were included for the study.

The exclusion criteria included malignancy history, surgery and/or trauma in facial, cervical and thoracic areas; severe cardiopulmonary, metabolic/endocrine diseases and other progressive or non-progressive central and peripheral neurologic disorders such as stroke, multiple sclerosis and cranial neuropathy,and patients who had insufficient consciousness state (subjects with the mini-mental test scores below 24).

Before the evaluation, the patients were given verbal and written information on the nature of the study. Informed consent forms were signed upon admission to the study. All procedures were conducted by the relevant principles of Helsinki Declaration. Also, approval of the study was obtained by the Gulhane School of Medicine Local Institutional Ethics Committee (allocated number: 342).

Demographic characteristics

Demographic and disease characteristics age, gender, disease's name anddisease duration (year)were recorded.

Instruments

All instruments were performed on all subjects within 72 hours.

Functional Oral Intake Scale (FOIS) (18)

The Functional Oral Intake Scale is an ordinal scale that is used to assess the feeding status and meaningful change in the oral intake. Oral intake status with this scale is graded from 1st level "nothing by mouth" to 7th level "total oral intake,no restrictions".

Fiberoptic Endoscopic Evaluation of Swallowing (FEES)

Endoscopic evaluation of patients was performed by the otolaryngology specialist using a 3.4-mm diameter non-ducted fiberoptic nasopharyngoscope (Karl Storz GmbH & Co KG, Tuttlingen, Germany) while the patient was in the vertical sitting position. The penetration and aspiration scale (PAS) determined by Rosenbek et al. (19) are leveled in 8 stage from stage 1 "material does not enter the airway" to stage 8 "material enters the airway, passes below the vocal folds, and no effort is made to eject" using 3 ml of water, a tablespoon of yogurt and a matchbox of bread.

Neuromuscular Disease Swallowing status Scale (NdSSS) (17)

For the NdSSS scoring, the patient's normal daily lunch at the hospital or at home was observed. The NdSSS consists of 8-stage scale. These 8 stages are from completely tube dependent state- Level 1 "Tube feeding with saliva suctioning in the oral cavity is necessary. A patient can neither discharge nor swallow saliva" to completely oral feeding- Level 8 "Totally orally fed with no restrictions. A patient eats all kinds of food".

High-Resolution Esophageal Manometry (HRM)

High resolution manometry (HRM) was performed using a 24-channel single use water-perfused catheter of 4.7 mm in diameter (Medical Measurement Systems, Netherlands) after 6 hours fasting. The catheter was inserted through the nasal cavity and advanced into gastric area with swallowing. Esophageal contraction capacity was evaluated by swallowing 10 times with 5 mL of water. Resting pressure was measured for 20 seconds. Computer analysis software (MMS software version 9.6, Netherlands) was used to assess various esophageal motility parameters and to perform standard analysis of the motor pattern according to the Chicago Classification version 3 (CCv3) (20).

Translation Procedures

Translation was performed according to the report of Beaton et al. (21).Permission to use and translate the questionnaire was obtained from the author Wada et al. (17). The NdSSSwas translated into Turkish by neurology, otolaryngology, gastroenterology and PMRspecialists independently. After comparing all translations and making necessary corrections, a Turkish version of the tool was created. It was thenback-translated into English in collaboration with a professional linguist. The final Turkish-NdSSS (T-NdSSS) was accepted following a comparison of the meaning and format with the original English form. After the pilot study was completed on 5 patients, the form was finalized by the feedback obtained.

Reliability

Inter-rater reliability between two raters were measured with Cohen's kappa (κ) and internal consistency was evaluated with the Cronbach's alpha (α) (22). Maximum 24 hours between the examinations was considered to be sufficient to prevent bias.

Validity

Convergent validity was determined by comparing the FOIS scores and concurrent validity was assessed by PAS (FEES) for oropharyngeal dysphagia as well asCCv3 (manometry) for esophageal dysphagia since there is no single test that evaluates both oropharyngeal and esophageal dysphagia. Endoscopic and manometric evaluation was performed by otolaryngology and gastroenterology specialists blinded to the design of the trial within the 72 hours after performing the T-NdSSS test.

Statistical Analysis

All statistical analyses were carried out by using SPSS 22.0 statistical package (SPSS, Chicago, IL, USA). Descriptive statistics were demonstrated as mean (standard deviations) and median for continuous variables and as a percentage (%) for nominal variables. $\chi 2$ test was used for differences among the disease groups in categorical variables. Cohen's κ and Cronbach's α coefficient for internal consistency were calculated as measures of the inter-rater reliability between the two raters.Cohen κ coefficients were rated as follows: 0.81-1.00 as excellent, 0.61-0.80 as very good, 0.41 0.60 as good, 0.21-0.40 as fair, and 0-0.20 as poor (22). A Cronbach's α>0.70 was considered acceptable (23). (For validity, Spearman's rank correlation test was used to indicate an correlation between FOIS, PAS and CCv3, and T-NdSSS levels. In addition, FOIS and PAS scores and CCv3 diagnoses were correlated with NdSSS levels for each NMD subgroups. The correlation coefficient (r) was used to show the power of correlation. According to this; <0.30 points indicated weak, 0.30 to 0.50 points indicated moderate,0.50 to 0.75 points indicated good correlation, and 0.75 to 1.0 point indicated a very good correlation between the variables (24). P<0.05 values were accepted as statistically significant.

Results

Individual and Group Scores

Patient Characteristics

The mean age of 115 patients was 58.18±17.62 years [79 (68.7%) female, 36 (31.3%) male]. Sixty-five (56.6%) patients had ALS, 32 (27.8%) had MG and 18 (15.6%) had myopathy. The mean of PAS score was 3.71±1.96 (median 4.00) and FOIS score was 5.71±1.85 (median 6.00). The majority of patients had PAS level 4 (Material enters the airway, contacts the vocal folds, and is ejected from the airway n=40, 34.8%), FOIS level 4 (total oral diet a single consistency n=37, 32.2%) and ineffective esophageal motility disorder (n=83, 72.2%). Demographic and disease characteristics of the patients

Table 1. — **Demographic, disease and dysphagia characteristics patients**

	n=115
Age (mean±SD)	58.18±17.62
Gender n(%)	
Female	79 (68.7)
Male	36 (31.3)
Disease n(%)	
ALS	65 (56.6)
MG	32 (27.8)
Myopathy	18 (15.6)
Disease duration (years)	3.46±1.45
PAS n(%)	
Level 1	27 (23.5)
Level 2	15 (13)
Level 3	10 (8.7)
Level 4	40 (34.8)
Level 5	4 (3.5)
Level 6	2 (1.7)
Level 7	16 (13.9)
Level 8	1 (0.9)
FOIS n(%)	
Level 1	0
Level 2	9 (7.8)
Level 3	16 (13.9)
Level 4	37 (32.2)
Level 5	13 (11.3)
Level 6	14 (12.2)
Level 7	26 (22.6)
CCv3 n (%)	
Achalasia	0
EGJ outflow obstruction	0
Absent contractility	5 (4.3)
Ineffective esophageal motility	83 (72.2)
Normal	27 (23.5)

SD: Standard deviation, ALS: Amyotrophic Lateral Sclerosis, MG: myastenia Gravis, PAS: penetration aspiration scale, FOIS: functional oral intake scale, CCv3: Chicago Classification version 3.

Table 2. — **Swallowing evaluation results according to the disease groups**

	ALS (n=65)	MG (n=32)	Myopathy (n=18)	p*		
				ALS-MG	ALS-Myo	MG-Myo
PAS n(%)						
Level 1	0	17 (53.1)	10 (55.6)			
Level 2	0	7 (21.9)	8 (44.4)			
Level 3	4 (6.1)	6 (18.7)	0			
Level 4	38 (58.5)	2 (6.3)	0	0.001	0.001	0.609
Level 5	4 (6.2)	0	0			
Level 6	2 (3.1)	0	0			
Level 7	16 (24.6)	0	0			
Level 8	1 (1.5)	0	0			
FOIS n(%)						
Level 1	0	0	0			
Level 2	5 (7.6)	2 (6.3)	2 (11.1)			
Level 3	16 (24.6)	0	0	0.001	0.001	0.518
Level 4	36 (55.4)	1 (3.1)	0			
Level 5	8 (12.3)	5 (15.6)	0			
Level 6	0	8 (25)	6 (33.3)			
Level 7	0	16 (50)	10 (55.6)			
CCv3 n (%)						
Achalasia	0	0	0			
EGJ outflow obstruction	0	0	0	0.002	0.001	0.732
Absent contractility	1 (1.5)	2 (6.3)	2 (11.1)			
Ineffective esophageal motility	64 (98.5)	15 (46.9)	4 (22.2)			
Normal	0	15 (46.9)	12 (66.7)			

ALS: Amyotrophic Lateral Sclerosis, MG: Myastenia Gravis, PAS: penetration aspiration scale, FOIS: functional oral intake scale, CCv3: Chicago Classification version 3. ***:** χ^2 test

Table 3. — **Inter-rater reliability for NdSSS levels**

1st Rater	**2nd Rater**								
	Level 1	**Level 2**	**Level 3**	**Level 4**	**Level 5**	**Level 6**	**Level 7**	**Level 8**	**Total**
Level 1	0	0	0	0	0	0	0	0	0
Level 2	0	16	0	0	0	0	0	0	16
Level 3	0	0	11	1	0	0	0	0	12
Level 4	0	0	0	12	0	0	0	0	12
Level 5	0	0	0	0	12	0	0	0	12
Level 6	0	0	0	0	0	11	1	0	12
Level 7	0	0	0	0	0	0	30	4	34
Level 8	0	0	0	0	0	0	4	13	17
Total	0	16	11	13	12	11	35	17	115

NdSSS: Neuromuscular disease swallowing status scale.

and comparison of swallowing evaluation results according to the disease groups are shown in Table 1 and 2. The swallowing evaluation results of the MG and myopathy groups were similar. Swallowing functions of the ALS patients were worse than the other two groups.

Summary of T-NdSSS

All patients were evaluated by both 1st and 2nd raters. No patients were identified as level 1. The 1st and 2nd raters detected level 2 in 16 (13.9%) patients and level 5 in 12 (10.4%) patients. While the 1st rater identified 12 (10.4%) patients for level 3, 4 and 6; 34 (29.6%) patients for level 7; and 17 (14.8%) patients for level 8.The 2nd rater identified 11 (9,6%) patients for level 3, 13 (11.3%) patients for level 4, 11 (9.6%) patients for level 6, 35 (30.4%)patients for level 7and 17 (14.8%) patients for level 8 (Table 3). The percentage of agreement between both raters was 100% for level 2,3,5,6, and 92.3% for level 4, 85.7% for level 7 and 76.5% for level 8. The floor effect was notobserved. The ceiling effect was acceptable (14.8%).

Reliability

Internal consistency

The NdSSS that performed by 1st and the 2nd raters indicated that the internal consistency was “acceptable” with a Cronbach’s α values of 0.993 and 0.987, respectively. When evaluated according to disease groups, Cronbach’s α coefficient was 0.996 and 0.992 for ALS, 0.823 and 0.819 forMG and, 0.882 and 0.879 for myopathy.

Inter-rater reliability

Inter-rater reliability κ coefficient was 0.907 (Table 3). Cohen’s κ value was 0.962 for ALS, 0.888 for MG

and 0.869 for myopathy. These values were "excellent" for ALS, myopathy and MG.

Validity

Convergent validity

A very good significant correlation was found between NdSSS and, the FOIS scores (Spearman's r for rater 1:0.927 and for rater 2:0.927). When evaluated separately according to disease groups, There are very good correlation between NdSSS levels and FOIS scores for ALS (Spearman's for rater 1:0.913 and for rater 2: 0.915) and good correlation for MG (Spearman's r for rater 1:0.508 and for rater 2: 0.521) as well as good correlation for myopathy (Spearman's r for rater 1: 0.511 and for rater 2: 0.581).

Concurrent validity

A very good significant correlation was found between NdSSS and PAS scores (Spearman's r for rater 1: -0.836 and for rater 2-0.841). Also, there was a positive good correlation between NdSSS and CCv3 diagnoses evaluating disorders of esophageal peristalsis (Spearman's r for rater 1: 0.677 and for rater 2: 0.679, p=0.001) (Table 4).

When evaluated separately according to disease groups, there was good correlation between NdSSS levels and PAS (Spearman's r for rater 1:-0.648 and for rater 2: -0.656); and CCv3(Spearman's r for rater 1:0.514 and for rater 2: 0.573) levels for ALS. For MG there was a good correlation between NdSSS levels; and CCv3 (Spearman's r for rater 1:0.577 and for rater 2: 0.622) and PAS (Spearman's r for rater 1: -0.504 and for rater 2: -0.519)scores. Also, for myopathy; a very good [CCv3 (Spearman's r for rater 1: 0.976 and for rater 2: 0.982)] and good [PAS(Spearman's r for rater 1: -0.516 and for rater 2: -0.550)] correlations were defined for myopathy.

Table 4. — **Spearman's rank correlation between NdSSS levels and swallowing results**

	NdSSS (1st rater) r(p)	NdSSS (2nd rater) r(p)
All subject (n=115)		
FOIS	0.927 (0.001)	0.927(0.001)
PAS	-0.841(0.001)	-0.836(0.001)
CCv3	0.679(0.001)	0.677(0.001)
ALS (n=65)		
FOIS	0.915(0.001)	0.913(0.001)
PAS	-0.656(0.001)	-0.648(0.001)
CCv3	0.573(0.037)	0.514(0.029)
MG (n=32)		
FOIS	0.521(0.016)	0.508(0.003)
PAS	-0.519(0.028)	-0.504(0.024)
CCv3	0.622(0.001)	0.577(0.001)
Myopathy (n=18)		
FOIS	0.581(0.011)	0.511(0.033)
PAS	-0.550(0.018)	-0.516(0.021)
CCv3	0.976(0.001)	0.982(0.001)

r: correlation coefficient, NdSSS: Neuromuscular disease swallowing status scale, PAS: penetration aspiration scale, FOIS: functional oral intake scale, CCv3: Chicago Classification version 3, ALS: Amyotrophic Lateral Sclerosis, MG: Myastenia Gravis.

Discussion

The present study aimed both to evaluate the applicability of the NdSSS for dysphagia in all phases of swallowing in various NMD patients and to investigate psychometric properties of Turkish version of the NdSSS. For this, internal consistency, inter-rater reliability, convergent validity and concurrent validity methods were used.

As a result of the study, the NdSSS was found applicable in detecting both oropharyngeal and esophageal dysphagia in patients with NMD. In addition, T-NdSSS was found to be reliable and valid for screening dysphagia in NMD patients.

Dysphagia is generally seen at a rate of 30-80% in adult NMD patients. This ratio can be as higher as 100% depending on the disease, disease progression and prognosis (2-4). In ALS, which is one of the most progressive forms of NMDs, the main cause of death is respiratory failure that is mainly caused by aspiration pneumonia (25,26). Although it was reported in previous studies that oral phase disorders such as weakness in tongue and lip movements and difficulty in chewing solid foods were observed in the early stages, and pharyngeal phase disorders that cause aspiration were observed in advanced stages of ALS (27,28), but recent studies have shown that the penetration and aspiration can occur in patients with oral impairment, even without bulbar involvement (25). An explanation for this observation is that since the swallowing is a synergistic behavior that requires coordination between muscles and involves sequential muscle movements,oral phase disorder, which is the first phase of swallowing, can have a domino effect on next phases of swallowing.

As the name implies, almost all NMDs affect the strength and coordination of orofacial muscles can lead to disrupted swallowing. Although the mechanism is different from that of ALS, oropharyngeal dysphagia may develop in myopathies and MG due to muscle weakness. (6,15,29). Moreover, esophageal dysphagia is also added to oropharyngeal swallowing disorders, especially in myopathies and MG patients (8,29). Regardless of the NMD subgroup, dysphagia affects the quality of life of patients and causes both nutritional deficiency and functional disability, as well as aspiration pneumonia and associated mortality increase (29,30). Therefore, early recognition of dysphagia and identification of the current swallowing state is essential to prevent possible complications and improve quality of life.(16,27). An ideal swallowing screening tool should be able to assess the severity of dysphagia in symptomatic patients and reveal the possible risk of dysphagia in asymptomatic patients. The important issue is to define the problem so that the rehabilitation program can be initiated, regardless of the progression rate of dysphagia or the

predominant involvement of the swallowing phase in patients with NMD. NdSSS is a screening test developed to define the swallowing status of the patient. It has not been translated into any other language so far.It has been used in studies involving patients with ALS, DMD, and spinal muscular atrophy and has been specified as the recommended scale in meta-analyzes (16,31-33). The original study (17) has been conducted in a total of 134 DMD and 84 ALS patients.The reliability assessment of the study has been performed in 50 patients with DMD and 84 ALS, and inter-rater reliability κ coefficient was 0.98 for ALS, 0.95 for DMD, indicating excellent inter-rater reliability. In the present study, although inter-rater reliability was particularly low for myopathy and MG patients, it was 0.90 for all patients, 0.96 for ALS, 0.88 for MG and 0.86 for myopathy patients, indicating excellent inter-rater reliability for allNMD patients and subgroups. Similar to the original study, NDSS and FOIS were found to be correlated for the entire patient group and ALS in the validation study. However, when evaluated according to the disease subgroups, the correlation coefficient was lower in the MG and myopathy groups, even thougether was no weak correlation. In the original study, a different PAS was applied using only semi-liquid and jelly with videofluoroscopy, in contrast we used 3 food consistency including liquid, semi-liquid and solid materials. Accordingly, we found an acceptable validity as good and very good in all NMDs and subgroups of NMDs.

The difference of our study from the original study is evaluation of swallowing with its all aspects, namely by including esophageal dysphagia.Therefore, MG and myopathy patients without severe OD were also included in the study. Interestingly, a good correlation was found between esophageal dysphagia assessed by manometry and NdSSS(Spearman's r for rater 1: 0.677 and for rater 2: 0.679, p=0.001). When this relationship was evaluated separately according to disease subgroups, an acceptable level of correlation in all subgroups and stronger correlation in myopathywas found.This result has two meanings. First, although NdSSS aims to measure an OD-based dysphagia state, as stated in the original study, it also gives us an idea about the esophageal dysphagia since the swallowing phases are intertwined with each other.Second, not only oropharyngeal dysphagia but also esophageal dysphagia should be evaluated in patients with NMD.This seems to be the case for primary muscle disease such as myopathy, neuromuscular junction disease such as MG, or motor neuron disease such as ALS.The esophagus which is composed ofentirely striated muscles is affected primarily or secondarily because of the nature of the NMDs(8,16,29).

The most important limitation of the present study was small size and the level 1 patients who are completely dependent on the tube was not included since the transport of severe patients for manometric evaluation was not possible. In addition, discriminative validity could not be performed because not all subgroups were available. Besides, patients with other types of NMDs such as muscular dystrophy and polyneuropathy could not be included in the study since there were no hospital admissions during enrolment.

In conclusion, NdSSS is a valid and reliable swallowing screening tool that can evaluate oro-pharyngo-esophageal dysphagia in NMD patients.

Conflict of interest

None.

Financial interest

None.

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