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CLINICAL RESEARCH ARTICLE

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Screening for oropharyngeal dysphagia in adult patients with neuromuscular diseases using the Sydney Swallow Questionnaire

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Abstract

Introduction/Aims: Oropharyngeal dysphagia is common in patients with neuromuscular diseases (NMDs). Its early recognition is vital for proper management. We tested a large cohort of adult NMD patients for oropharyngeal dysphagia using the Sydney Swallow Questionnaire (SSQ). We also looked for possible differences in characteristics of oropharyngeal dysphagia in various NMD groups and diseases. Finally, we compared results of this screening with those from their corresponding medical records for eventual "clinical history" of dysphagia.

Methods: We asked patients to fill in the SSQ during follow-up outpatient visits at our neuromuscular reference center. A total score above the cutoff score of 118.5 out of 1700 was indicative of oropharyngeal dysphagia.

Results: Of the 304 adult patients assessed for eligibility, 201 NMD patients (96 women and 105 men, aged 49.0 ± 16.2 years) were included and tested in this study. Oropharyngeal dysphagia was detected in 45% of all the NMD patients when using the SSQ, whereas only 12% had a positive medical record for dysphagia. The median SSQ scores for patients with myotonic syndromes (including myotonic dystrophy type 1), with amyotrophic lateral sclerosis, and with facioscapulohumeral dystrophy were above the cutoff score. The SSQ scores obtained revealed distinct oropharyngeal dysphagia characteristics in the different NMD groups and diseases.

Discussion: The SSQ tests positively for oropharyngeal dysphagia in a higher proportion of NMD patients compared with their medical records. The distinct oropharyngeal dysphagia characteristics we revealed in different NMD groups and diseases may help to elaborate adapted clinical approaches in the management of oropharyngeal dysphagia.

KEYWORDS
neuromuscular diseases, oropharyngeal dysphagia, screening, Sydney Swallow Questionnaire

Abbreviations: ALS, amyotrophic lateral sclerosis; CMT, Charcot-Marie-Tooth disease; DM1, myotonic dystrophy type 1; FSHD, facioscapulohumeral dystrophy; HMSN, hereditary motor and sensory neuropathies; NMD, neuromuscular disease; PPS, postpolio syndrome; QoL, quality of life; SSQ, Sydney Swallow Questionnaire; SSQ(+), patients with a SSQ score above the cutoff of 118.5; VFSS, videofluoroscopic swallow study.

1 | INTRODUCTION

Oropharyngeal dysphagia is common in patients with neuromuscular diseases (NMDs). It can be defined as impairment affecting the transit and airway-protective functions of the oropharyngeal structures, namely the upper esophageal sphincter, pharynx, larynx, tongue, or mouth, in isolation or combination.[1-3] Its prevalence varies from 34% to 80% in NMD patients.[4-8] Oropharyngeal dysphagia can appear early in the course of these diseases and may lead to malnutrition, dehydration, aspiration pneumonia, or difficulty in managing secretions.[9-11] It may also have social and psychological consequences, with a negative impact on the patient's quality of life (QoL).[12] Oropharyngeal dysphagia is sometimes difficult to diagnose and this can delay initiation of treatment. Some NMD patients may be unaware of their oropharyngeal dysphagia (silent aspiration) and manifestations may be nonspecific (pneumonia, weight loss, or cough).[6,13]

To minimize related complications, appropriate management is essential. This involves early detection and referral for appropriate clinical evaluation and follow-up.[14,15] As such, systematic screening could contribute to the early management of oropharyngeal dysphagia in NMDs.[9,16] An ideal screening tool should have high sensitivity and specificity.[17] Among existing tools, the Sydney Swallow Questionnaire (SSQ) meets all these criteria and is specifically designed to detect oropharyngeal dysphagia.[16,18] In addition to being available and validated in French, previous studies have demonstrated its content, construct, discriminant and predictive validity, and its test-retest reliability to detect oropharyngeal dysphagia in a range of different disorders.[16,19-24] Although it has been used to evaluate patients with Duchenne muscular dystrophy (DMD), we found no data regarding its use in the assessment of oropharyngeal dysphagia in other NMDs.

The main aim of this study was to determine the proportion of patients identified as having oropharyngeal dysphagia by the SSQ in a large cohort of adult NMD patients and to compare the results of this testing with those from their medical records, namely regarding any mention of dysphagia. We also looked for differences in the characteristics of oropharyngeal dysphagia in different NMD groups and diseases.

2 | METHODS

2.1 | Participants and ethical considerations

We recruited all patients between August 2017 and May 2018 during their routine outpatient visits to the neuromuscular reference center of the Cliniques universitaires Saint-Luc (Brussels, Belgium). All patients gave written informed consent in accordance with the Declaration of Helsinki and the current guidelines for Good Clinical Practice. Inclusion criteria were a diagnosis of NMD, age between 18 and 80 years, and the ability to speak and read French. Exclusion criteria were inability to complete the questionnaire, recent history of surgery (≤1 month), and having other progressive disease(s) in addition to NMD. Patients without a definitive diagnoses were excluded. The study was approved by the institutional medical ethics committee (B403201628760) and registered on clinicaltrial.gov (NCT02845362).

2.2 | Assessment means

The SSQ is a patient self-reporting questionnaire. It consists of 17 questions with a maximum total score of 1700. Each question (except question 12 [Q12]) is scored using a visual analog scale (100 mm), anchored at each end by extreme statements.[16,22] Q12 is scored 0 to 5, based on the duration of an average meal, and then multiplied by 20. Total score is calculated by summing the scores for each question.[16] Average completion time is approximately 5 minutes.[16,20,24] In this study, the validated French version of the SSQ and the cutoff score of 118.5 (sensitivity of 93% and specificity of 82%), defined during the validation of this French version, were used in our NMD population.[24] A total score above the cutoff value was considered indicative of oropharyngeal dysphagia. The higher the score, the greater the risk of oropharyngeal dysphagia.

2.3 | Protocols and data collection

Each participant completed the SSQ during a follow-up outpatient visit at the neuromuscular reference center. Oral and written explanations summarizing the purpose of the study were provided by the investigator. Medical records were reviewed to look for information related to the eventual presence of reported dysphagia based on all swallowing assessment tools other than the SSQ. The different diseases in our cohort were sorted into different groups based on the classification of the 2019 version of the Gene Table of Neuromuscular Disorders (Table S1).[25]

2.4 | Statistical analysis

Descriptive analysis was performed for demographic characteristics. Normality of the distribution was verified using the Kolmogorov-Smirnov test. Depending on the normality of the distribution, data were expressed as mean ± standard deviation or as median (interquartile range), and parametric or nonparametric tests were used when appropriate. The results for the different groups and diseases were compared using the Kruskal-Wallis test. We used Pearson's correlation coefficient (r) for calculating correlations. The r values range from $+1$ to -1 and the absolute value qualifies the strength of the correlation as negligible ($0.00 < r < 0.09$), weak ($0.10 < r < 0.39$), moderate ($0.40 < r < 0.69$), strong ($0.70 < r < 0.89$), and very strong ($r > 0.90$).[26] The coefficients of variation were calculated for the different groups and diseases (standard deviation divided by the mean). Finally, the phi (ρ) coefficient was used to verify the associations between the different data and was reported according to Yule.[27] The ρ coefficient ranges from -1 to $+1$, with negative numbers representing negative relationships, zero representing no relationship,

and positive numbers representing positive relationships.[27] Data analysis was carried out only for groups and diseases with more than 10 subjects using SPSS version 25.0 (IBM Corp, Armonk, New York). $P \leq .05$ was considered significant.

3 | RESULTS

3.1 | Patients' characteristics

A total of 304 patients were assessed for eligibility. Of these, 201 met the inclusion and exclusion criteria and were included in the final analysis (96 women and 105 men, aged 49.0 ± 16.2 years). Figure 1 is a flowchart of the study. There were 11 different NMD groups and 29 different diseases represented (Table 1). The characteristics of the most represented NMD groups and diseases are highlighted in Table 2.

3.2 | Screening for oropharyngeal dysphagia and SSQ scoring

Oropharyngeal dysphagia was detected in 45% of all the NMD patients using the SSQ. All SSQ scores and the percentages of patients who tested positive (ie, attained an SSQ score above the cutoff score of 118.5; SSQ[+]) are presented in Table 2. The median total SSQ score was higher than the cutoff score in myotonic syndromes, amyotrophic lateral sclerosis (ALS), myotonic dystrophy type 1 (DM1), and facioscapulohumeral dystrophy (FSHD) patients. The coefficients of variation calculated for the different groups and diseases ranged from 9% to 13%.

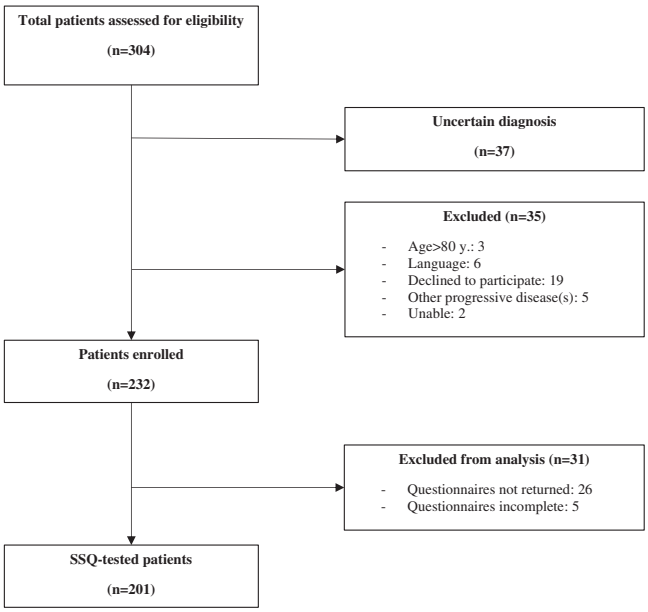

FIGURE 1 Flow diagram of the study (N = 201). Abbreviations: SSQ, Sydney Swallow Questionnaire; y, years

3.3 | Characteristics of oropharyngeal dysphagia assessed by SSQ for different NMD groups and diseases

The scores for each question and their correlations with the total score are presented in Tables S2 and S3. In patients with muscular dystrophies, these correlations were strong for Q9 (residues) and weak for Q13 (regurgitation and coughing during meals). For the other questions, correlations were moderate. For myotonic syndromes, the questions related to food types or consistencies were strongly correlated, regardless of bolus consistency. Questions related to timing (Q12 and Q14), nasal regurgitation (Q13), and QoL and enjoyment (Q16 and Q17) did not correlate significantly with the total score. In

TABLE 1 Distribution of patients (neuromuscular disease groups and diseases)

Groups	n	Recruited patients
Group 1: Muscular dystrophies	44	21.9%
Duchenne and Becker muscular dystrophies	19	
Facioscapulohumeral dystrophy	19	
Others[a]	6	
Group 6: Myotonic syndromes	24	11.9%
Myotonic dystrophy type 1	23	
Others[a]	1	
Group 12: Spinal muscular atrophy and motor neuron diseases	29	14.4%
Amyotrophic lateral sclerosis	19	
Others[a]	10	
Group 14: Hereditary motor and sensory neuropathies	41	20.4%
Charcot-Marie-Tooth disease	32	
Hereditary spastic paraplegia	5	
Others[a]	4	
Group 16: Other neuromuscular disorders	43	21.4%
Postpolio syndrome	15	
Chronic inflammatory demyelinating polyneuropathy	7	
Others[a]	21	
Other groups	20	10.0%
Group 2: Congenital muscular dystrophies	2	
Group 3: Congenital myopathies	4	
Group 4: Distal myopathies	4	
Group 5: Other myopathies	2	
Group 9: Metabolic myopathies	6	
Group 11: Congenital myasthenic syndromes	2	

[a]Less than five subjects per pathology.

TABLE 2 Characteristics of the most represented (>10 subjects) NMD groups and diseases

	n	Age[a] (years)	Total SSQ score[b]	SSQ(+) (%)	Patients with "medical records" for dysphagia
Total	201	49.0 ± 16.2	88 (33-257)	45.3%	12.4%
Muscular dystrophies	44	44.5 ± 15.8	112 (36-273)	47.7%	4.5%
Myotonic syndromes	24	42.2 ± 14.3	168 (64-320)	66.7%	24.5%
Spinal muscular atrophy and motor neuron diseases	29	54.0 ± 15.5	79 (24-321)	41.4%	37.9%
Hereditary and sensory neuropathies	41	44.6 ± 15.7	60 (24-140)	31.7%	2.4%
ALS	19	56.9 ± 12.8	177 (20-326)	52.6%	47.4%
DMD/BMD	19	33.2 ± 11.7	64 (35-312)	42.1%	5.3%
CMT	32	45.6 ± 14.8	66 (28-169)	37.5%	3.1%
DM1	23	42.3 ± 14.6	173 (75-328)	69.6%	26.1%
FSHD	19	54.5 ± 11.2	136 (40-271)	52.6%	5.3%
PPS	15	62.1 ± 8.4	61 (23-184)	26.7%	0.0%

Abbreviations: ALS, amyotrophic lateral sclerosis; CMT, Charcot-Marie-Tooth disease; DM1, myotonic dystrophy type 1; DMD/BMD, Duchenne and Becker muscular dystrophy; FSHD, facioscapulohumeral dystrophy; NMD, neuromuscular disease; PPS, postpolio syndrome; SSQ, Sydney Swallow Questionnaire; SSQ(+), patients with a score above the cutoff of 118.5.

[a]Data expressed as mean ± standard deviation.

[b]Data expressed as median (interquartile range).

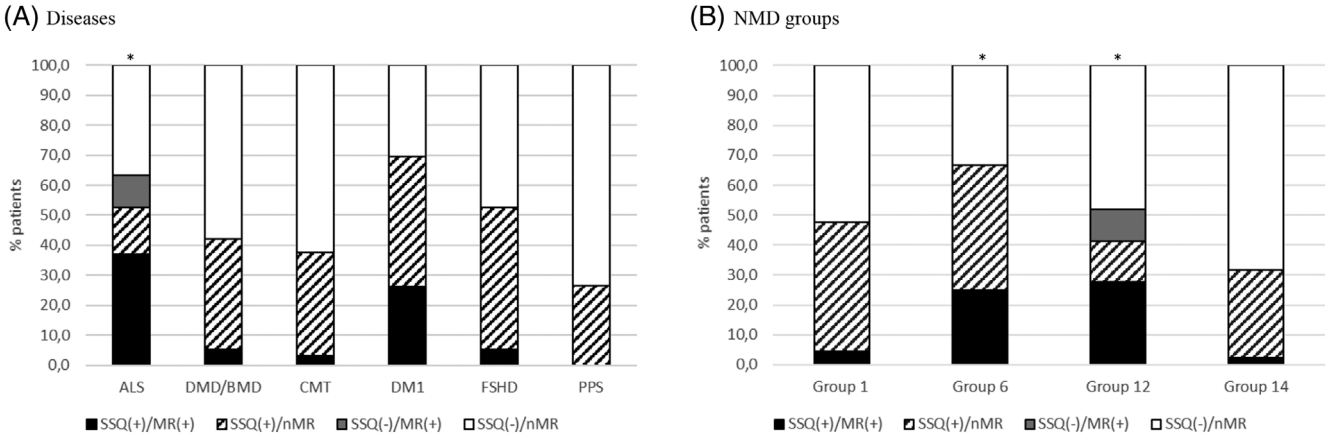

FIGURE 2 Comparison between dysphagia results from medical reports and results of our screening tool for oropharyngeal dysphagia (SSQ score). Group 1: Muscular dystrophies. Group 6: Myotonic syndromes. Group 12: Spinal muscular atrophy and motor neuron diseases. Group 14: Hereditary and sensory neuropathies. Asterisk indicates positive and significant relationship. All other relationships are not significant. Abbreviations: ALS, amyotrophic lateral sclerosis; CMT, Charcot-Marie-Tooth disease; DM1, myotonic dystrophy type 1; DMD/BMD, Duchenne and Becker muscular dystrophies; FSHD, facioscapulohumeral dystrophy; MR(+), patients with a positive medical record for dysphagia; NMD, neuromuscular disease; nMR, patients without any medical record for dysphagia; PPS, postpolio syndrome; SSQ, Sydney Swallow Questionnaire; SSQ(+), patients with a score above the cutoff of 118.5; SSQ(−), patients with a score under the cutoff of 118.5

spinal muscular atrophy and motor neuron diseases (SMA/MNDs), the scores of most of the questions (59%) correlated strongly with the total score. Only the correlation of Q16 (severity of dysphagia) was weak. Finally, in hereditary motor and sensory neuropathies (HMSNs), the score for questions linked to swallowing difficulties for soft food or saliva (Q4 and Q7), starting a swallow (Q8), and time to eat (Q12) did not correlate significantly with the total score. The other *r* values ranged from 0.334 to 0.636 (weak to moderate correlations).

With regard to specific disorders, correlations were very strong for Q10 and Q11 for patients with Duchenne and Becker muscular dystrophies, linked to coughing and choking. Except for Q15 (regurgitation and coughing during meals), other correlations were moderate to strong (*r* from 0.457 to 0.898). In FSHD, more than 85% of the question scores were at least moderately correlated (*r* from 0.427 to 0.837). In ALS, the questions related to food types or consistencies were strongly correlated, regardless of bolus consistency (*r* from 0.739 to 0.865). In Charcot-Marie-Tooth disease (CMT), the correlations were strong for Q8 (starting a swallow), Q9 (residues), and Q10 (coughing with solids).

3.4 | "Clinical history" of dysphagia as per medical records

The percentage of patients in each NMD group or disease who had been identified as having dysphagia in their medical records (ie, dysphagia based on assessments other than SSQ) is illustrated in Table 2. Among the 201 patients, only 25 had a positive medical record for dysphagia. The proportion of patients with positive medical records for dysphagia was significantly different between NMD groups ($P < .001$). Proportions of patients with positive medical records for dysphagia were the highest in myotonic syndromes and SMA/MNDs.

Regarding individual diseases, the proportions of patients with a medical record for dysphagia were also significantly different ($P < .001$). The proportion of patients with a medical report of dysphagia in DM1 and ALS were 26% and 47%, respectively, whereas all other diseases were below 6%. Notably, none of the postpolio syndrome (PPS) patients had a positive medical record for dysphagia.

3.5 | Comparison between dysphagia results from medical records and results of our screening tool for oropharyngeal dysphagia

Among the entire NMD cohort analyzed, only 24% of the SSQ(+) patients had a positive medical record for dysphagia. Conversely, 97% of patients who tested negative for oropharyngeal dysphagia with the SSQ cutoff score had no medical record for dysphagia (Figure 2). A significant and moderately positive relationship between medical record of dysphagia and SSQ(+) was observed in the entire cohort ($\rho = 0.324$; $P < .01$). The relationships were significant and strongly positive for myotonic syndromes ($\rho = 0.408$; $P < .05$), SMA/MNDs ($\rho = 0.498$; $P < .01$), and ALS ($\rho = 0.478$; $P < .05$). All other relationships were not significant. Among the patients with medical record of dysphagia ($n = 25$), 21 had previously been assessed for oropharyngeal dysphagia by instrumental examinations and all were found positive. These instrumentations included fiber-optic endoscopic evaluation of swallowing (17 patients), videofluoroscopic swallow study (VFSS) (3 patients), and VFSS with manometry (1 patient). Among the 21 patients, 20 were SSQ(+) and 1 had a score of 107.

4 | DISCUSSION

Forty-five percent of NMD patients included in this study were found to test positive for oropharyngeal dysphagia using the SSQ, whereas only 12% had a positive medical record for dysphagia. Furthermore, SSQ scoring revealed varying characteristics of oropharyngeal dysphagia in the different NMD groups and diseases.

4.1 | Muscular dystrophies

In Duchenne and Becker muscular dystrophies, Q9 (residues), Q10 (coughing with solids), Q14 (more than one swallow), and Q16 (severity of dysphagia) scored higher than the other questions. This may correspond to the swallowing problems (well known in these types of dystrophies) linked to facial weakness, impaired mastication, poor coordination of the tongue, and incomplete pharyngeal clearance of solid food in these patients.[5,9,28-31] In FSHD, questions related to swallowing hard foods and residues (Q5 and Q9) scored high. Weakness of the orofacial muscles, especially the tongue, would induce a delayed oral- and pharyngeal phase in FSHD.[32-34]

4.2 | Myotonic syndromes

All patients in this group except one had DM1. The scores were high for most of the questions, regardless of bolus consistency. Swallowing impairments in DM1 are linked to facial and oral weakness and problems of coordination between swallowing and breathing. Patients need more chewing cycles to prepare bolus and yet present more residues.[35-37] Discrepancies between SSQ total score and answers to Q16 (severity of dysphagia) and Q17 (impact on the patient's QoL) were observed. This could be related to reported influences of mood or insight issues on the filling of self-reported questionnaires in DM1 patients.[38]

4.3 | Spinal muscular atrophy and motor neuron diseases

The scores for this group were higher than for the other groups for all questions, except those related to cough (Q10, Q11, and Q15), nasal regurgitation (Q13), and food stuck in the throat (Q9). In these patients, all phases of swallowing are affected and lead to increased risks of aspiration.[39,40] Andersen et al also showed that involvement of respiratory muscles limits coughing, leading to secretion accumulation, lung infections, and eventual respiratory failure.[41] Given the absence of sensory impairment in SMA/MND patients, the results of Q9 should be related to other physiopathological imperfections. There is also impaired cough reflex with age and disease progression.[42,43] Oropharyngeal dysphagia is very common in ALS patients and involves alterations in forming and holding the bolus, irrespective of its consistency.[44,45] In our results, the scores for this disease were the highest for almost all questions. Only Q15 (regurgitation and coughing during meals) did not contribute significantly to the total score.

4.4 | Hereditary motor and sensory neuropathies

In this group, all the SSQ(+) patients fell in the CMT subgroup. Surprisingly, oropharyngeal involvement is rarely reported in CMT.[46] The scores were higher for questions not related to food types or consistencies, namely Q16 (severity of dysphagia) and Q17 (impact on the patient's QoL). Underlying reasons are not known but the mindset of patients could be implicated. Indeed, CMT patients exhibit an increased tendency for depressive symptoms compared with the

general population.[47] However, oropharyngeal dysphagia could be part of a subclinical neurogenic dysfunction, which could be an alternative contributory cause. Further clinical evaluation with formal swallowing assessment tools would therefore be necessary for further studies.

4.5 | Postpolio syndrome

In PPS, the scores for Q9 (residues) and Q11 (coughing with liquids) were higher than for the other questions. Oropharyngeal dysphagia may develop during the initial (acute) phase of the viral infection or may occur after several years.[48,49] Subclinical bulbar innervated muscle dysfunction could underlie a progressive weakness in oropharyngeal and laryngeal muscles.[50,51] This weakness could lead to muscle fatigue during eating, and could be difficult to identify by instrumental examinations.[48,49,51,52]

In all NMD groups and diseases analyzed, the median score for Q12 (time to eat) was 20, which is not considered as clinically relevant as our preceding results revealed that Q12 rarely rated zero even in healthy subjects.[24]

Regarding the sensitivity and characteristics of the screening tools in detecting oropharyngeal dysphagia, the following observations were notable: Many patients with no indication of dysphagia in their medical record were positive for oropharyngeal dysphagia using the SSQ. The number of SSQ(+) patients in whom oropharyngeal dysphagia had not been identified previously (based on their medical records) was substantial in PPS, FSHD, and other muscular dystrophies.

The choice of the cutoff score to define oropharyngeal dysphagia in this study was crucial as it seemingly permitted the detection of a high proportion of patients with swallowing impairments. In previous studies using the SSQ, the different cutoff scores for defining dysphagia ranged from 111.0 to 234.0.[16,19-24] As such, the SSQ assessment tool appears to be an easy and effective method for detecting oropharyngeal dysphagia in NMDs. Thus, the SSQ could effectively become a first-step approach in a detection strategy that may ultimately be supplemented by further eventual confirmatory diagnostic tools such as instrumentations/interventional procedures.

Medical records notably showed that the number of patients with detected dysphagia diagnosed with methods other than SSQ was highest in patients with myotonic syndromes, ALS, and DM1.

Some diseases may appear to be underrepresented in this cohort, but their rates remain concordant with those reported in data from the Belgian Neuromuscular Disease Registry.[53] It may also be noteworthy that variables such as diagnostic delay, respiratory function, medications, diet modifications, and clinical assessment of bulbar function were not taken into consideration in this study. They could vary greatly from patient to patient, and over time. This could modify the scoring results accordingly. Follow-up SSQ rescoring could thus provide further insights based on evolution or treatment.

Finally, an eventual categorization of adult NMDs on the basis of the varied characteristics of dysphagia we found in different adult NMDs would be of interest as it would complement that establish by van den Engel-Hoek et al for pediatric NMDs.[37]

In conclusion, our study has revealed that a high proportion of adult NMD patients tested positive for oropharyngeal dysphagia using the SSQ compared with results derived from their medical records. Awareness of this outstanding observation could impact clinical management of NMD patients. Finally, distinct oropharyngeal dysphagia characteristics were found among different NMD groups and diseases; this may permit the development of adapted clinical approaches in the management of oropharyngeal dysphagia in these patients.

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CONFLICT OF INTEREST

None of the authors have no conflicts of interest to declare.

ETHICAL PUBLICATION STATEMENT

We confirm that we have read the Journal's position on issues involved in ethical issues in publications and affirm that this report is consistent with those guidelines.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of this article.

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