Screening and evaluation tools of dysphagia in adults with neuromuscular diseases: a systematic review

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Abstract

Background: The purpose of this systematic review was to summarize the different dysphagia screening and evaluation tools, and to identify their measurement properties in adults with neuromuscular diseases (NMDs).

Methods: A systematic review was performed based on the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. The search strategy was conducted across three databases (*PubMed, CINAHL* and *ScienceDirect*). Measurement properties of each tools and the Quality Index, developed by Downs and Black, were considered for the different investigated studies.

Results: The search strategy produced 2221 articles. After removal of duplicates and full-text analysis, 19 studies were included. Most of the publications focused on amyotrophic lateral sclerosis (ALS; $n = 10$) and Duchenne muscular dystrophy (DMD; $n = 4$). A total of 12 tools, listed as instrumental and noninstrumental examinations, were retrieved. A total of five of them used videofluoroscopic swallow study (VFSS). Measurement properties of the tools are not completely described in detail in many studies. The neuromuscular disease swallowing status scale, a noninstrumental tool, is the only one that assessed all measurement properties in ALS patients. The median score reported for the Quality Index was 16.

Conclusions: This systematic review identified 12 different tools for the screening and evaluation of dysphagia in adults with NMD. Majority of the studies presented VFSS as a valid and reliable examination to assess dysphagia in ALS and DMD. Other tools were mainly evaluated in ALS patients, but further studies are needed to complete their measurement properties. In other NMDs, no firm conclusion can be made because of insufficient data and heterogeneity of NMDs.

Keywords: dysphagia, neuromuscular diseases, screening, evaluation, tools, amyotrophic lateral sclerosis, impaired swallowing

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Introduction

Impaired swallowing, also called dysphagia, is one of the most critical problems in patients with neuromuscular diseases (NMDs) and can be related to increased morbidity and mortality.[1,2] In adult patients with NMDs, dysphagia is present in 34.9–80%,[1,3–10] depending on several factors including the genetic mutation, symptoms, age at onset, rate of progression, and prognosis.[2,11,12] Early signs related to dysphagia, such as 'wet voice', silent aspiration, or loss of weight, are often discreet and unclear[13,14] and overall prevalence and incidence are challenging due also to the lack of standardized assessment procedures.[1–5,9] Specific disorders such as bulbar and progressive respiratory muscle weakness, often associated with NMDs, disrupt the ability to swallow safely and efficiently and may lead to severe complications, such as malnutrition, dehydration, aspiration pneumonia, and other pulmonary sequelae.[2,5,6,10,15,16] Therefore, assessment of swallowing problems appears to be a high priority for NMD patient's caregivers. Dysphagia detection should contribute to earlier management, and possible prevention, of comorbidities, including the impact on

Table 1. The PICOS (participant, intervention/exposure, comparator, outcome and study design).

Criterion	Description
Population	Patients with neuromuscular diseases. Adult patients.
Intervention	Validation of one or more tools for screening or evaluation of dysphagia.
Comparison	Comparison between a minimum of two tools.
Outcomes	Psychometrics properties, characteristics of each tool.
Study design	All research studies not classified as a review or meta-analysis.

quality of life.[2,17–19] Previous guidelines and studies considered fiberoptic endoscopic evaluation of swallowing (FEES) or videofluoroscopic swallowing study (VFSS) as the standard criteria for the evaluation of swallowing problems in adult patients with non-NMD disorders.[20–23] However, such evidence does not exist in adult patients with NMDs and different assessment strategies are utilized depending on the center, the country and the usual practices.[2,20,24,25] International guidelines concerning patients with NMDs only mentioned that patients with a clinical history of swallowing difficulties or recurrent chest infections should have a specialist assessment by a speech and language therapist (SLT) if the swallow is thought to be unsafe.[24,25] An ideal swallowing assessment tool should offer quantitative measures and be indicated for patients with NMDs with symptoms or underlying conditions potentially associated with dysphagia to eliminate or minimize the complications of dysphagia.[2,26,27] For this evaluation, it also appears important to quantify the severity and progression of dysphagia. For asymptomatic patients at risk of dysphagia, screening tools need to detect early symptoms or swallowing abnormalities and identify those that need more definitive swallowing assessment.[28,29] Adequate rehabilitation and quantification of treatment efficiency for clinical purposes or research is possible with both. They indicate a precise cutoff score and, ideally, be cost-effective, easy to interpret and not too time-consuming.[3,19,27,28,30–32] The purpose of this systematic review was to summarize the different dysphagia screening and evaluation tools, and to identify their measurement properties in adults with neuromuscular diseases.

Materials and methods

The Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines were followed during the stages of design, analysis, and reporting of this systematic review.[33,34] The protocol has been registered in PROSPERO (Registration No. CRD42016033690). The research strategy followed the same pattern and the same criteria as our previous systematic review in children with NMDs.[35] The full search strategy is highlighted in Supplement 1. Online databases were screened from inception to June 2018. The PICOS (participant, intervention/exposure, comparator, outcome and study design) approach was applied for data extraction (Table 1). After removing duplicates, abstracts were selected based on relevance by two independent investigators (N.A. and G.R.). Full-text articles were assessed when inclusion was uncertain from the title and abstract. Where there was disagreement, a consensus meeting was organized to determine eligibility. Articles were excluded if they included insufficient information on the instrument used. Study details and data were extracted by N.A. and G.R. Data extracted included the name of the tests, sample characteristics (including sample size, age group and disease severity), test protocols, outcomes, and correlations. Measurement properties of investigated tools, defined following the COSMIN statement, were reported when available and were described in two categories: 'instrumental' and 'noninstrumental' examinations.[36] As described by Mann, we classified the different publications as cohort, cross-sectional, or case-control studies.[37] The Quality Index, developed by Downs and Black for assessing methodological quality and bias, was applied by the two same investigators.[38,39] This tool covers 27 questions relating to the study description and external and internal validity, with a total maximum score of 28.[40] Each study was assigned a grade of 'excellent' (24–28 points), 'good' (19–23 points), 'fair' (14–18 points) or 'poor' (<14 points).[40]

Results

A total of 2221 references were retrieved in the different databases (Figure 1). After removal of

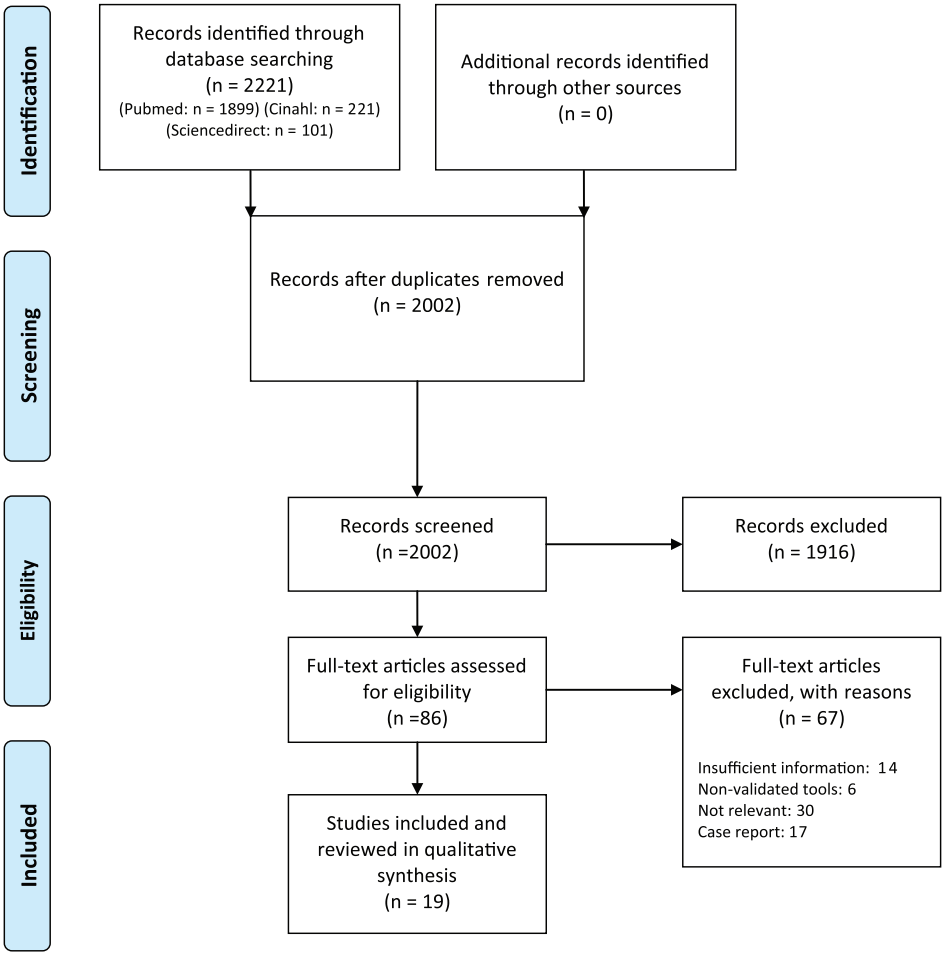

Figure 1. Systematic review flow diagram.

duplicates, and full-text analysis, 19 studies met the inclusion criteria of this systematic review.

All the studies were published from 1997 to 2017. Out of the 19 included studies, 6 were Japanese. The other were conducted in European countries (10), USA (2) and Turkey (1). All the studies used observational research methods. A total of four involved adult and child patients in their studied samples.[41–44] Studies characteristics are summarized in Table 2. The Downs and Black index ranged from 12 to 19 with a median total score of 16. Most studies were classified as 'fair'.[7,10,14,41–43,45–54] Two were assigned as 'poor'[55,56] and only one study presented a 'good' quality index[44] (Table 3).

Table 4 summarizes the different tools used to study dysphagia for each separate NMD. Out of the 19 studies, 10 focused on amyotrophic lateral sclerosis (ALS), and 4 on Duchenne muscular dystrophy (DMD). Other pathologies studied included myotonic dystrophy type 1 (DM1), inclusion body myositis (IBM), myasthenia gravis (MG), spinal muscular atrophy (SMA), polymyositis/dermatomyositis (PM/DM), Friedreich's ataxia (FA) and spinal and bulbar muscular atrophy (SBMA). With regards to the tools, most publications (n = 5) used VFSS.[10,42,48,55,56] Surface electromyography[14,43,54] (sEMG) and FEES[10,45] were found in three and two studies, respectively. Other tools listed with measurement properties and characteristics are listed in Supplement 2.

Instrumental examinations

Validity

The VFSS, also reported as the modified barium swallowing examination, is the most commonly assessed tool in NMDs for dysphagia assessment[10,42,48,55,56] or as reference to compare the other techniques.[7,41,46,49–54] In ALS, two generic

Table 2. Characteristics of included studies ($n = 19$).

Study	Study design	Number of cases ($n = 1.101$)	Number of controls	Underlying diseases
Cosentino and colleagues[57]	CSS	26	30	ALS
Hiraoka and colleagues[56]	CSS	25	n/a	ALS
Plowman and colleagues[58]	CSS	70	n/a	ALS
Plowman and colleagues[53]	CSS	70	n/a	ALS
Olthoff and colleagues[54]	CCS	20	n/a	IBM
Wada and colleagues[44]	CSS	218	n/a	ALS, DMD
Murono and colleagues[59]	CSS	19	n/a	ALS
Pilz and colleagues[48]	CS	45	10	DM1
Aydogdu and colleagues[14]	CS	364	297	ALS, DM1, MG, PM/DM
Mano and colleagues[55]	CSS	47	38	SBMA
Archer and colleagues[47]	CSS	15	12	DMD
Archer and colleagues[46]	CSS	15	12	DMD
Paris and colleagues[49]	CCS	20	n/a	ALS
Cox and colleagues[7]	CSS	43	n/a	IBM
Hanayama and colleagues[45]	CSS	31	n/a	DMD
Higo and colleagues[60]	CCS	11	n/a	MG
Kidney and colleagues[51]	CSS	25	n/a	ALS
Briani and colleagues[10]	CSS	23	n/a	ALS, SMA
Mari and colleagues[50]	CS	14	n/a	ALS, DM1, FA

ALS, amyotrophic lateral sclerosis; CCS, case-control study; CS, cohort study; CSS, cross-sectional study; DM1, myotonic dystrophy type 1; DMD, Duchenne muscular dystrophy; FA, Friedreich's ataxia; IBM, inclusion body myositis; MG, myasthenia gravis; n/a, not available; PM/DM, polymyositis/dermatomyositis; SBMA, Spinal and bulbar muscular atrophy; SMA, spinal muscular atrophy.

scales were used for VFSS analysis, the dysphagia outcome severity scale (DOSS)[57] and the penetration aspiration score (PAS).[48,59] Murono and colleagues suggested that evaluation of swallowing kinematics is one of the major advantages of VFSS.[55] In patients with ALS, they observed that the oral phase seemed the most affected and pharyngeal contraction was correlated with PAS and may play a role in penetration or aspiration even in patients without bulbar symptoms ($p < 0.01$). However, they showed that aspiration or penetration is not common in those patients in the first stages of the disease.[55] In Briani and colleagues, VFSS findings were compared with FEES and manometry findings.[10] VFSS had a significantly greater sensitivity (92%), to highlight swallowing impairment, especially when silent in ALS and SMA patients.[10] In contrast, all of the nondysphagic patients also showed radiological swallowing abnormalities indicating a specificity of 0% in this study.[10] In DMD patients, Hanayama and colleagues suggested that VFSS abnormalities were related to advanced age, except for impaired oral holding.[42] VFSS is a better indicator for the oral phase of swallowing ($p < 0.05$) and the pooling of contrast fluid in the valleculae ($p < 0.05$) than the 'dysphagia questionnaire', a standard set of questions related to the frequency of 10 symptoms of upper gastrointestinal dysfunction.[42,60] In MG, the abnormal laryngeal elevation observed during VFSS was

Table 3. Quality Index developed by Downs and Black. Scores for each included study.

Study	Reporting	External validity	Bias	Confounding	Power	Total	Grades+
	(11)*	(3)*	(7)*	(6)*	(1)*	(28)*	
Cosentino and colleagues[57]	8	1	3	2	0	14	Fair
Hiraoka and colleagues[56]	6	1	3	2	0	14	Fair
Plowman and colleagues[58]	8	1	5	2	0	16	Fair
Plowman and colleagues[53]	8	1	5	2	0	16	Fair
Olthoff and colleagues[54]	8	0	5	2	0	15	Fair
Wada and colleagues[44]	8	2	5	3	0	18	Fair
Murono and colleagues[59]	6	0	5	2	0	13	Poor
Pilz and colleagues[48]	9	1	5	1	0	16	Fair
Aydogdu and colleagues[14]	8	1	4	3	0	16	Fair
Mano and colleagues[55]	9	1	3	3	1	17	Fair
Archer and colleagues[47]	9	1	6	3	0	19	Good
Archer and colleagues[46]	8	1	5	2	0	16	Fair
Paris and colleagues[49]	10	0	5	1	0	16	Fair
Cox and colleagues[7]	6	3	4	2	0	15	Fair
Hanayama and colleagues[45]	7	3	5	1	0	16	Fair
Higo and colleagues[60]	6	1	4	1	0	12	Poor
Kidney and colleagues[51]	7	1	5	2	0	15	Fair
Briani and colleagues[10]	10	0	5	2	0	17	Fair
Mari and colleagues[50]	6	3	6	2	0	17	Fair

*Maximum score that can be given for each item with the Quality Index developed by Downs and Black.
+Grading maximal score with Quality Index developed by Downs and Black: Excellent (24–28 points); Good (19–23 points); Fair (14–18 points); Poor (<14 points).

significantly correlated with aspiration ($p = 0.001$) and may lead to predicting aspiration and pneumonia.[56] However, across the different studies evaluating VFSS, a great variety of methodological settings was observed in terms of thickness, viscosity and volume of contrast fluids used (Table 5).

Manometry was used in patients with ALS and SMA and showed that an abnormal upper esophageal sphincter (UES) opening and hypotonia of the proximal pharynx were the most sensitive physiological signs of dysphagia (80%).[10] When compared with controls, the patients of this study presented a greater number of incomplete UES openings and a significantly extended 'intra-bolus' pharyngeal pressure.[10] As demonstrated in VFSS, dysphagia appeared to be linked to the presence of a defective oropharyngeal phase of swallowing. It should be noted that the specificity of pharyngo-esophageal manometry is weak in this study (20%).[10] According to Briani and colleagues, FEES examination was not sensitive in highlighting swallowing alterations both in dysphagic and in nondysphagic patients (53%) but had a good specificity to rule out other organic causes of dysphagia (85%).[10]

Teams from the United Kingdom (UK), Turkey and Italy used sEMG to assess swallowing and

Table 4. Tools used to study dysphagia in each NMD.

Underlying diseases	Tools	Study
ALS	VFSS, sEMG (DL), FEES, V-VST, Man., VCA, 3SwT, NdSSS, MTP, EAT-10	Murono and colleagues[55]; Aydogdu and colleagues[14]; Mari and colleagues[47]; Paris and colleagues[46]; Plowman and colleagues[49,50,62]; Kidney and colleagues[48]; Wada and colleagues[41]; Briani and colleagues[10]; Cosentino and colleagues[54]; Hiraoka and colleagues[53]; Plowman and colleagues[49,50,62]
DMD	VFSS, SSQ, sEMG, NdSSS	Archer and colleagues[43]; Archer and colleagues[44]; Hanayama and colleagues[42]; Wada and colleagues[41]
DM1	FEES, sEMG (DL), 3SwT	Pilz and colleagues[45]; Aydogdu and colleagues[14]; Mari and colleagues[47]
MG	sEMG (DL), VFSS	Higo and colleagues[56]; Aydogdu and colleagues[14]
IBM	Standard Questionnaire, RT-MRI	Cox and colleagues[7]; Olthoff and colleagues[51]
SMA	VFSS, Man., FEES	Briani and colleagues[10]
PM/DM	sEMG (DL)	Aydogdu and colleagues[14]
FA	3SwT	Mari and colleagues[47]
SBMA	MTP	Mano and colleagues[52]

3SwT, 3-ounce water swallow test; ALS, amyotrophic lateral sclerosis; DL, dysphagia limit; DM1, myotonic dystrophy type 1; DMD, Duchene muscular dystrophy; EAT-10, eating assessment tool; FA, Friedreich's ataxia; FEES, fiberoptic endoscopic evaluation of swallowing; IBM, inclusion body myositis; Man., pharyngo-esophageal manometry; MG, myasthenia gravis; MTP, maximum tongue pressure; NdSSS, neuromuscular disease swallowing status scale; NMD, neuromuscular disease; PM/DM, polymyositis/dermatomyositis; RT-MRI, real-time magnetic resonance imaging; SBMA, spinal and bulbar muscular atrophy; sEMG, surface electromyography; SMA, spinal muscular atrophy; SSQ, Sydney Swallow Questionnaire; VCA, voluntary cough airflow; VFSS, videofluoroscopic swallow study; V-VST, volume-viscosity swallow test.

dysphagia in patients with DMD, ALS, DM1, PM/DM and MG.[14,43,54] Aydogdu and colleagues used the dysphagia limit (DL) to compare the peak duration and relative timing of muscle activity during swallowing of four muscle groups (orbicularis oris, masseter, submental, and infrahyoid muscles) in ALS, DM1, PM/DM and MG patients.[14] DL is a quantitative and noninvasive method for the assessment of swallowing impairment. It represents the volume at which two or more swallows become necessary to swallow the whole bolus. Values for DL were compared with the clinical degrees of dysphagia, graded from 1 (no clinical signs and symptoms of dysphagia) to 4 (obvious clinical signs and symptoms of dysphagia, including aspiration).[61] Patients with clinical dysphagia had abnormal DL for <20 ml of water.[14] DL was significantly correlated with the improvement or worsening of dysphagia for ALS, PM/DM and MG ($p < 0.001$) but not for DM1 patients ($p > 0.05$). Also, DL was useful to assess the effect of treatment as well as changes of dysphagia over time.[14] Archer and colleagues tested peak activity of four muscle groups during swallowing between DMD patients with dysphagia, those with preserved muscle function, and healthy controls.[43] Although there were no differences in the timing or duration of muscle activity between the three groups, these comparative data provided insight into the relative activity of the orbicularis oris, masseter, infrahyoid, and submental muscles during swallowing. Thus, a distinction between a preserved and disordered swallowing function in patients with DMD could be made.[43] Compared with controls, patients with DMD had to use a greater maximal muscle activity to swallow [masseter ($p < 0.01$), orbicularis oris ($p < 0.05$), submental ($p < 0.05$)], indicating muscle weakness. However, no differences were found between DMD with dysphagia and those patients with an intact swallowing function.[43]

Table 5. Protocols used in VFSS studies.

Articles	Consistencies/food	Frequency/bolus	Criteria used for diagnosing dysphagia
Cox and colleagues[7]	n/a	0, 3, 6 or 9 ml of opaque fluid	Signs subdivided in two categories: IP or AR.
Hanayama and colleagues[45]	**Liquid:** 70% water + 30% Omnipaque **Jelly swallow:** strawberry jelly + 30% of volume Omnipaque **Solid:** Barium pancakes (60% flour/40% barium powder)	**Liquid:** 1 × 30 ml **Jelly swallow:** 3 × 5 ml **Solid:** 1 piece of cake cut in 3 pieces and put into the patient's mouth	Impaired oral hold, oral residuals, pooling in the valleculae, pooling in the valleculae after repeated swallow, pooling in the pyriform sinus, pooling in the pyriform sinus after repeated swallow, supraglottic penetration, pharyngo-oral regurgitation.
Higo and colleagues[60]	'Appropriate and safe bolus textures' (thin liquid or semisolid)	1, 3, 5 ml or self-regulated	Bolus transport from the mouth to the pharynx, bolus holding in the oral cavity, velopharyngeal seal, tongue base movement, pharyngeal constriction, laryngeal elevation, upper esophageal sphincter opening, and bolus stasis at the pyriform sinus + lung aspiration.
Murono and colleagues[59]	140% barium mixture + other contrast and consistencies (n/a)	3 ml	All 15 physiologic components except for a component of esophageal clearance, proposed by Martin-Harris and Jones, were thoroughly evaluated from the perspective of all six oral components and all eight pharyngeal components.
Kidney and colleagues[51]	Varied food from liquid to solid	n/a	PAS, DOSS.
Hiraoka and colleagues[56]	Yogurt (282 mPas) containing contrasting agent	3 g	Qualitative evaluation: (1). Tongue function, (2). Residue accumulation; Quantitative evaluation (1). Bolus formation and oral transit time (second): (2). Pharyngeal transit time (second).
Plowman and colleagues[58]	**Liquid:** Varibar thin barium sulphate suspension, (EZ-EM, Inc., Westbury, NY, USA). **Pudding:** EZ-pudding, (EZ-EM, Inc.). **Thin liquid:** n/a	A standardized bolus presentation protocol: (1). two 1-cc boluses of liquid contrast; (2). one 3-cc of thin liquid contrast; (3). one 3-cc of pudding; (4). one 20-cc bolus of liquid contrast; (5). 90 cc sequential swallows of thin liquid contrast; (6). in the anterior-posterior view, the patient was administered a 20-cc bolus of liquid contrast.	PAS
Plowman and colleagues[53]	Idem as Plowman and colleagues[58]	Idem as Plowman and colleagues[58]	Idem as Plowman and colleagues.[58]
Olthoff and colleagues[54]	Liquid contrast agent (either Imeron 350 or Gastrolux)	1 × 20 ml	Relevant parameters of swallowing included bolus control and transport, velo-pharyngeal closure, laryngeal penetration, aspiration, and bolus retention in the pharyngeal tract.
Mano and colleagues[55]	40% weight per volume barium sulphate	3 ml	PAS + residue quantification.
Briani and colleagues[10]	Fluid/Semisolid	Chosen by the patient	Oral stasis; loss of barium in the mouth vestibula during swallowing; repetitive tongue movements; incomplete or inadequate velopharyngeal closure; presence of aspiration; functionality of upper esophageal sphincter and lower esophageal sphincter; pharyngo-esophageal motility.

AR, aspiration-related dysphagia; cc, cubic centimeter; DOSS, dysphagia outcome severity scale; IP, dysphagia due to impaired propulsion; mPas, millipascal seconds; n/a, not available; PAS, penetration and aspiration scale.

In 2017, Cosentino and colleagues investigated electrophysiologically submental/suprahyoid activity (SHEMG) and laryngeal-pharyngeal movements (LPMs) during swallowing in patients with ALS ($p = 0.004$).[54] They detected reduced SHEMG and LPM in patients with dysphagic ALS. A strong to moderate correlation was observed between these two indices and the PAS and DOSS score, respectively. Moreover, a decrease in swallowing reproducibility could be a preclinical sign of dysphagia and, beyond a certain threshold, a pathological hallmark of oropharyngeal dysphagia. Interestingly, SHEMG was the only electrophysiological parameter correlated with the disease duration and could be an expression of the progressive degeneration of the motor neurons occurring in the course of the disease ($r = 0.494$, $p = 0.010$).[54]

Overall, two studies assessed the validity of maximum tongue pressure (MTP) in ALS and SBMA patients.[52,53] Both used a digital tongue pressure manometer equipped with a balloon probe (JMS Co. Ltd., Hiroshima, Japan) and asked the participants to compress the balloon upward onto their palates for 7 seconds, three times at 1-minute intervals, using the maximum voluntary effort of the tongue. The maximum value of these three measurements was considered as the MTP for each patient.[52,53] Hiraoka and colleagues evaluated the relationship between MTP and the characteristics of swallowing disorders in patients with ALS.[53] They showed that MTP was significantly lower in the patients with ALS with reduced tongue function ($p = 0.002$) or with pharyngeal residue ($p = 0.006$) than in the patients with normal characteristics. Bolus formation and oral and pharyngeal transit time observed in VFSS were significantly prolonged among those with reduced MTP ($p < 0.01$).[53] In SBMA, Mano and colleagues described a decrease in patients within 3 years from the onset of the disease compared with healthy controls ($p < 0.001$). In comparison with VFSS, the values of tongue pressure in the patients with laryngeal penetration were significantly smaller than those of the patients without ($p = 0.018$).[52]

Real-time magnetic resonance imaging (RT-MRI) allowed precise time measurements and identification of the respective tissue morphology.[51] In 20 patients, penetration was seen in 10% and 30% of them by RT-MRI and FEES or VFSS, respectively. Bolus retention in the pharyngeal tract is believed to be the most sensitive indicator of functional deficits in swallowing and was reliably identified by all three modalities (FEES, VF, RT-MRI). The Bland–Altman plots did not reveal any systematic deviation among those methods.[51]

Finally, Plowman and colleagues identified the cough volume acceleration (CVA), peak expiratory flow rate (PEFR) and peak expiratory flow rise time (PEFRT) as significant predictors of penetration/aspiration status in 70 ALS patients.[62] Following this study, ALS patients with a CVA less than 45.3 l/s^2 were 5.6 times more likely to penetrate/aspirate. A PEFR lower than 4.0 l and a PEFRT greater than 80 ms increased 3.6 and 3.2 times the risk of penetration/aspiration, respectively.[62] Those cutoffs for CVA, PEFR and PEFRT had sensitivities of 91.3%, 82.6%, and 73.9% respectively and, specificities of 82.2%, 73.9%, and 78.3% for identifying ALS penetrator/aspirators.[62]

Reliability

Reliability was evaluated in five studies focused on five different NMD populations. Kidney and colleagues investigated inter- and intra-rater reliability of VFSS in relation to the DOSS and PAS, and analysis of videofluoroscopy was undertaken in ALS patients. These results indicated acceptable test–retest reliability when using the DOSS and the Aspiration-Penetration Rating Scale (APRS) with inter-rater reliability of 92% and 95% of agreement, respectively and intra-rater reliability scored at 98% and 100% of agreement, respectively.[48] Pilz and colleagues described inter- and intra-rater reliability as sufficient for all FEES variables (piecemeal deglutition, delayed initiation pharyngeal reflex, post-swallow vallecular and pyriform pooling, laryngeal penetration or tracheal aspiration), with a weighted kappa > 0.61 in DM1 patients.[45] For all FEES variables, bolus consistency significantly influenced the likelihood of observing mild or severe swallowing impairment: patients were more likely to have impaired swallowing with thin liquids than with thickened fluids (odds ratio > 1).[45] Archer and colleagues have suggested that physiological changes in the swallowing muscles activity in DMD can be distinguished between preserved and disordered swallowing function with sEMG.[43] They repeated the sEMG procedure in order to examine the reproducibility of the results. The mean differences between the repeated sEMG assessments were amplitude: 29.12 ± 21.69% MVC, duration: 0.69 ± 0.62 s, and relative timing: 0.40 ± 0.24 s. Mano and colleagues performed the

test–retest of MTP in 24 patients with SBMA on two different occasions at an interval of 21.3 ± 3.9 days. They found no statistically significant difference. The intraclass correlation coefficient was 0.986 ($p < 0.001$), indicating an excellent test–retest reliability of the tongue pressure. The RT-MRI was compared with FEES and VFSS in a cohort of patients with IBM.[51] The inter-rater agreement was assessed for bolus transport and retention. The Krippendorff α was 0.39 (RT-MRI), 0.55 (VF), and 0.67 (FEES) for bolus transport, and 0.51 (RT-MRI), 0.52 (VFSS), and 0.52 (FEES) for retention.[51]

Responsiveness

Responsiveness has not been evaluated in studies concerning instrumental examinations.

Noninstrumental examination

Validity

Validity was assessed in five studies about noninstrumental examinations.

Plowman and colleagues compared the eating assessment tool 10 (EAT-10), a 10-question self-administered, symptom-specific dysphagia outcome tool to score patients with ALS.[50] The total EAT-10 score was calculated with scores ranging from 0 (no impairment) to 40 (severe impairment) and demonstrated good discriminant ability to accurately identify ALS penetrator/aspirators (PAS ⩾ 3) with a cutoff score of 3 [area under the curve (AUC): 0.77, sensitivity: 88%, specificity: 57%].[63] It demonstrated excellent accuracy at identifying aspirators (PAS ⩾ 6) utilizing a cutoff score of 8 (AUC: 0.88, sensitivity: 86%, specificity: 72%). On average, EAT-10 scores were five times higher in ALS aspirators than in those patients who demonstrated safe swallowing.[50] The neuromuscular disease swallowing status scale (NdSSS), an 8-stage scale for dysphagia in patients with progressive NMD, was correlated with PAS significantly in patients with ALS ($r = -0.51$) but in DMD patients ($r = 0.22$).[41] Another examination, the volume-viscosity swallow test (V-VST) showed a high sensitivity and specificity (93% and 80%, respectively) for screening and diagnosing oropharyngeal dysphagia in patients with ALS.[46] The V-VST involved administering increasing fluid volumes of different textures/thicknesses and the assessment of efficiency and safety of swallowing.

In DMD patients, Archer and colleagues showed that the Sydney swallow questionnaire (SSQ),[19] a self-administered questionnaire based on 17 questions using visual analogue scales, detected dysphagia with good sensitivity and specificity (78 and 83%, respectively) at a cutoff score of 234 (total maximum score = 1700).[44] In this study, the dysphagic status of DMD participants were defined with a clinical history of dysphagia and the functional oral intake score. The 3-ounce water swallow test (3SwT), validated previously in other patient groups (e.g. stroke, head and neck surgery, Parkinson's disease, dementia),[58,64,65] was compared with VFSS and a 25-item form (one of them is 'history of cough'), identifying dysphagia. The 3Swt showed a higher specificity than clinical signs (86 *versus* 30%) but a lower sensitivity (52%) compared with VFSS in patients with NMD.[47] The association of 'history of cough' (coughing episode reported by the patient) with the 3SwT gave an increase in both positive and negative predictive values (84 and 76%, respectively).[47]

Finally, Cox and colleagues assessed impaired food propulsion or aspiration-related problems in patients with IBM by comparison of a standard questionnaire, previously validated in patients with Parkinson's disease,[66] with VFSS results.[7] The authors showed that the questionnaire had a better sensitivity and positive predictive value for impaired food propulsion than for aspiration-related problems (70 *versus* 63% and 92 *versus* 65%, respectively) and that two questions reliably predicted the presence of impaired food propulsion on VFSS, namely 'Does food get stuck in your throat?' and 'Do you have to swallow repeatedly in order to get rid of food?'. However, dysphagia was more frequently designated by VFSS when used as a reference than the standard questionnaire results (79 *versus* 65%).[7]

Reliability

Inter- and intra-rater reliabilities were only assessed for the NdSSS, in 50 patients with DMD and 84 patients with ALS. For inter- and intra-rater reliabilities, the weighted kappas were 0.95 and 1.00, respectively, for DMD; and 0.98 and 0.98, respectively, for ALS.[41]

Responsiveness

Responsiveness was determined with the standardized response mean for the NdSSS and was large in ALS (1.21) and moderate in DMD (0.65).[67]

Discussion

The purpose of this systematic review was to summarize the different dysphagia screening and evaluation tools and to identify their measurement properties in adults with NMDs. A total of 19 articles were identified based on strong criteria. Evaluation tools were mainly found for patients with ALS with eight different tools.[10,14,41,46–50,53–55] Validity has been assessed in the majority of the studies in instrumental and noninstrumental examinations even if specificity and sensibility were frequently lacking. Reliability has been verified for VFSS, FEES, sEMG, MTP, RT-MRI and NdSSS but was incomplete mainly in noninstrumental examinations.[41,43,45,48,51,52] Responsiveness has been reported only for NdSSS in patients with ALS and DMD.[42]

VFSS was used as primary outcome in 5 studies [10,42,48,55,56] and as comparator in 10 out of the retrieved studies.[7,41,46,49–54] Among them, we observed a great heterogeneity in the VFSS procedures and analysis used as highlighted in Table 5. This disparity was already found in prior reviews of swallowing assessment in other diseases.[20,31,32,68,69] In addition, previous studies reported poor inter-rater reliability for VFSS, depending on the procedure and bolus consistency used to assess swallowing in adult patients.[70,71] Equally, five reviews in non-NMD adult patients emphasized that consensus on the terminology and study protocols (e.g. fluid thickness used during studies) is needed for more useful comparison with VFSS,[31,32,69,72] like for sEMG or FEES.[14,43]

The importance of bolus properties was highlighted by Ciccero and colleagues[73] They insisted on the considerable changes in physiology observed depending of the bolus consistency (volume, viscosity, solid or fluid), size, method of ingestion (e.g. cup, straw) and chemosensory input (taste, smell, sensory receptors) in the normal swallow.[73] Those properties need to be considered by the clinician during dysphagia assessments when considering what is normal from what is pathological.[73,74] Classically, patients with solid food dysphagia are more likely to have disorders of esophageal phase, whereas those with difficulty with liquids are more likely to have oropharyngeal dysphagia.[75] However, in NMD patients, this dichotomy may be artificial because it is well known that those with oropharyngeal dysphagia can have dysphagia for liquids and solids in the different phases of swallowing, specific patterns of dysphagia depending on the underlying disease.[2,3,75] For example, DMD patients may have difficulties with chewing and oropharyngeal transport of solid foods, as well as pharyngeal residue without aspiration is more common and is likely due to muscle weakness.[2] In ALS, difficulties may likely be inability to hold bolus, reduced mastication, residue in the oral cavity and delayed swallow reflex.[76] Moreover, all patients with NMDs may also have difficulty with management of excessive thick mucus, which may contribute to breathing discomfort. The use of bolus with different properties seems also extremely helpful in the assessment of patients with NMDs in order to assess the further therapeutic and dietetic management. In the different studies included, those data were lacking, neither for cutoff level nor standardization are present specifically for the different NMDs.

Several details need to be highlighted about instrumental examinations. First, the fatigability, an important factor in patients with NMDs with muscle weakness, was generally not taken into account during the assessments.[4,77] We can hypothesize that tools only give a snapshot and do not consider factors such as fatigability or ventilatory support needed.[4,44] Secondly, some assessments are invasive and can cause discomfort to the patient (e.g. manometry and FEES), and potentially involves exposure to radiation (e.g. in VFSS and RT-MRI). Finally, examinations that include large volumes of liquids may put the patient at risk of aspiration and choking.[27,44] In addition, VFSS, MRI or FEES are often not possible in daily routine or in out-of-hospital settings such as physiotherapist and SLT practices due to the required expertise, the patient compliance, and the evolution of the swallowing disorder which can be sometimes rapidly progressive.[29] In regular follow-up assessments, even if the specific equipment needed may be considerate expensive,[2] the cost burden is often distributed among departments (radiology, gastroenterology, or otolaryngology) and implementation can be only limited in underdeveloped countries.[31]

Noninstrumental examinations, done subjectively or by self-administration, can be interesting alternatives for screening, follow up or complementation of instrumental evaluations in the diagnosis. These may be useful, in particular, in the assessment of the fluctuating nature of the motor and emotional symptoms in NMDs and their impact on quality of life.[78]

From a clinical point of view, following the definition outlined previously,[3,19,27,28,30–32] we can

consider that noninstrumental examinations (SSQ, V-VST, EAT-10 and 3SwT), voluntary cough airflow (VCA) and sEMG represent ideal screening or follow-up tools with high sensitivity and specificity and are quick, cost-effective, and easy to interpret.[28] Moreover, they give accurate cutoff scores and are more effective to detect early swallowing problems or risk factors. Instrumental examinations (VFSS and FEES) showed good validity and should be used for diagnosis or when aspiration status is unclear from noninvasive approaches. Following previous studies, VFSS also presented an important role in the follow up of aspiration occurrence and the evaluation of the improvements related to the treatment, and maybe the reduction of symptoms. On the other side, FEES can provide real-time visual feedback during swallowing therapy contrary to VFSS. FEES may be helpful to test many therapeutic interventions and many strategies without a time limit when the patient's intake and quality of life depend on a diet upgrade. FEES allows the assessment of a meal in a functional, real-life situation. Fatigue increases can also be checked as the meal progresses, but also if positioning, rate of intake, and method of feeding impact the safety. Moreover, the study of Andersen and colleagues has shown the interest of FEES in the assessment of the laryngeal responses during treatment with mechanical insufflation-exsufflation.[79]

Further studies are needed to standardize assessment procedures (tools, settings and chronology) according to the evolution and the specificities of each adult patient with an NMD. Moreover, to evaluate deglutition in patients with NMDs, minimal clinically important differences, which reflect changes related to clinical interventions, could improve those different examinations.

It would also be relevant to adapt tools to the timing of occurrence and evolution of the disease. Indeed, dysphagia may vary with the natural course of the disease and either be present from the early stages of the disease progression or appear with time in end-stage patients. For example, Andrenelli and colleagues showed that patients with ALS and dysphagia had a global dysfunction of the oral and pharyngeal swallowing phases with more difficulties for swallowing thin liquids when conversely, DM1 patients with dysphagia were younger, tended to obesity and had greater muscular impairment than those without dysphagia and will have difficulties in swallowing a solid bolus.[16] In another way, dysphagia in DMD worsens with age, with increasing mastication effort.[80] Recognition of specific clinical profiles for the different disease and stage will support and guide the detection of swallowing disorders in patients with NMDs. Furthermore, with new treatments (like noninvasive ventilation), life expectancy of patients with NMDs continues to increase. It will be more and more important in the future to deal with associated complications such as swallowing impairments and treatment such as noninvasive ventilation or airway clearance technique.[1,12,81,82]

The main limitation of this systematic review was the quality level of the included studies. Indeed, most of them were rated as 'fair' and we have noted the absence of randomized controlled trials. Also, some methodological issues need to be discussed. As observed by Olthoff and colleagues, the thickness and volume of the bolus used is sometimes different in the evaluation of the tools in the same study and firm conclusion must be made carefully.[51] Also, the positioning of the patients during the different assessments was poorly documented and is a source of bias. Most of the assessment is in a posture which does not reflect actual conditions of swallowing in daily life. The impact of positioning and head control has been demonstrated to be an important parameter.[83] Only Hanayama and colleagues and Hiraoka and colleagues evaluated the patients while seated in their own wheelchairs.[42,53] The characteristics and measurement properties of the tools are not always described in detail in many studies (Supplement 2), and it is often difficult to distinguish among screening, evaluation and diagnosis.[69,84] Frequently, the objectives of the different tools were not clearly defined by the different authors. Screening is defined as the presumptive identification of unrecognized disease in an apparently asymptomatic population by means of tests, examinations or other procedures that can be applied rapidly and easily to the target population.[31,85,86] This screening can be done by questionnaires, observations, physical evidence, among others.[87] Validated self-reported questionnaires have been increasingly used in clinical research as well as clinical practice to capture the individual's perspective regarding their disease symptoms.[88] When identified by the screening tool, the patient should be referred for diagnosis of swallowing disorders, conducted from clinical evaluation and supplemented, when necessary. Diagnosis aims to determine presence of dysphagia, its severity and changes that it may cause, and the rehabilitation plan.

Conclusion

This systematic review identified 12 different tools for the screening and evaluation of dysphagia in adults with NMDs. Among them, NdSSS, a non-instrumental tool, is the only one who assessed all measurement properties in patients with ALS. The majority of the studies presented VFSS as a valid and reliable examination to assess dysphagia in patients with ALS and DMD. Other tools were mainly evaluated in patients with ALS, but further studies are needed to complete their measurement properties. In other NMDs, no firm conclusion can be made because of insufficient data and heterogeneity of NMDs. Recognition of specific clinical profiles for the different disease and stage will support and guide the detection of swallowing disorders. Each tool presents specific measurement properties to be more designed for diagnosis or screening and could be used appropriately according to the evolution and the specificities of each adult patient with an NMD.

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