Résumé

Plus de 30% des personnes atteintes de sclérose en plaques (paSEP) souffrent de troubles de la déglutition, ce qui représente un taux plus élevé que supposé précédemment. La dysphagie neurogénique (DN) peut être à l'origine de différents types de troubles sensori-moteurs de l'oropharynx chez les paSEP et s'avère liée à la fois au degré de handicap et aux signes d'atteinte du tronc cérébral. Environ 15% des paSEP dont le handicap est léger peuvent également souffrir de DN. Les outils diagnostiques comprennent l'anamnèse, l'examen de dépistage au chevet (test de 50 ml d'eau avec évaluation de la sensation pharyngée ou oxymétrie de pouls) et parfois une étude de radioscopie télévisée de la déglutition (ERTD) et une évaluation fibroscopique de la déglutition (EFD). L'ERTD et l'EFD sont des méthodes complémentaires qui présentent chacune des avantages et des inconvénients. Les interventions pour le traitement de la DN chez les paSEP sont principalement basées sur la thérapie de déglutition fonctionnelle, comprenant des méthodes de restitution, de compensation et d'adaptation. L'objectif de l'intervention consiste en la prévention de l'aspiration et de la pneumonie par aspiration. L'évaluation des résultats doit porter sur des paramètres cliniquement pertinents tels que la diminution et la restriction des activités, ainsi que la qualité de vie.

Überblick

Mehr als 30% der Personen mit multipler Sklerose (PmMS) leiden an Schluckstörungen. Dies ist ein höherer Prozentsatz als bisher angenommen. Neurogene Dysphagie (ND) kann verschiedene oropharyngeale Störungen der Sensormotorik bei PmMS bewirken und wird sowohl mit dem Behinderungsgrad als auch mit Hirnstammzeichen in Verbindung gebracht. Ungefähr 15% der PmMS mit geringer Behinderung können ebenfalls an ND leiden. Diagnostische Methoden umfassen die Aufnahme der Krankengeschichte, Bedside-Screening-Tests (50-ml-Wasser-Test kombiniert mit einer Bewertung der pharyngealen Empfindung oder mit Pulsoxymetrie) und gegebenenfalls eine videofluoroskopische Schluckstudie (VFSS) oder eine fiberoptische endoskopische Bewertung des Schluckens (FEBS). VFSS und FEBS sind komplementäre Methoden, und beide haben Vor- und Nachteile. Interventionen für ND bei PmMS basieren hauptsächlich auf funktioneller Schlucktherapie, einschließlich Methoden zur Restitution, Kompensation und Adaption. Ziel der Interventionen ist, der Aspiration und Aspirationspneumonie vorzubeugen. Die Bewertung der Ergebnisse sollte sich auf klinisch relevante Parameter konzentrieren wie z. B. Einschränkung der Aktivitäten, Erschwernis der Teilnahme und gesundheitsbedingte Lebensqualität.

Sommario

Oltre il 30% dei pazienti affetti da sclerosi multipla (SM) soffre di sintomi che interessano la deglutizione, una percentuale più alta di quanto si supponesse in precedenza. La disfagia neurogena (DN) può causare nella SM molti tipi diversi di disfunzioni motorie sensoriali orofaringee, ed è associata sia al grado di disabilità sia ai segni del tronco cerebrale. Può soffrire di DN anche circa il 15% dei casi di SM con leggera disabilità. Gli strumenti diagnostici includono la valutazione dell'anamnesi remota del paziente, un semplice esame di screening (test di 50 ml d'acqua abbinato alla valutazione della sensazione faringea o all'ossimetria del polso), a volte uno studio video radioscopico della deglutizione (SVRD) e la valutazione endoscopica mediante fibre ottiche della deglutizione (VEFOD). Lo SVRD e la VEFOD sono metodi complementari ed offrono sia vantaggi che svantaggi. Gli interventi sulla DN dovuta alla SM si basano principalmente sulla terapia funzionale della deglutizione, includendo metodi di restituzione, compensazione ed adattamento. Lo scopo dell'intervento è prevenire l'aspirazione e la polmonite ab ingestis. La valutazione dei risultati dovrebbe concentrarsi sui parametri clinicamente rilevanti, quali limitazione dell'attività, restrizione della partecipazione e qualità di vita connessa alle condizioni di salute.

Reseña

Más del 30% de las personas con esclerosis múltiple tienen síntomas derivados de alteraciones de la deglución, una cifra mayor que el porcentaje supuesto previamente. La disfagia neurógena puede ocasionar muchos tipos diferentes de disfunciones sensitivomotoras orofaríngeas en las personas con esclerosis múltiple, y está asociada con el grado de disfunción neurológica y la presencia de signos de afectación del tronco del encéfalo. Aproximadamente el 15% de las personas con esclerosis múltiple con una leve disfunción neurológica pueden sufrir también de disfagia neurógena. Las herramientas de diagnóstico comprenden la obtención de la historia clínica, la exploración clínica selectiva (Administración de 50 ml de agua combinada con la evaluación de la sensación faríngea o con la pulsoximetría) y en ocasiones un estudio videofluoroscópico de la deglución y una evaluación endoscópica por medio de fibra óptica de la deglución. Estos dos últimos estudios son complementarios y ambos tienen ventajas e inconvenientes. Las intervenciones en la disfagia neurógena en los pacientes con esclerosis múltiple, se basan principalmente en el tratamiento de la deglución funcional, que incluye métodos de restitución, compensación y adaptación. El objetivo de la intervención es evitar la aspiración y la neumonía por aspiración. Las evaluaciones de los resultados se deben centrar en parámetros clínicamente relevantes, como la limitación de la actividad, la restricción de la participación y la calidad de vida relacionada con la salud.

Dysphagia and Multiple Sclerosis

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Summary

Over 30% of persons with multiple sclerosis (pwMS) suffer from swallowing symptoms, a higher rate than previously assumed. Neurogenic dysphagia (ND) may cause many different kinds of oropharyngeal sensorimotor dysfunctions in pwMS, and is associated with both the amount of disability and brainstem signs. About 15% of pwMS with mild disability may also suffer from ND. Diagnostic tools comprise history taking, bedside screening examination (50 ml water test combined with assessment of pharyngeal sensation or with pulse oximetry) and sometimes a videofluoroscopic swallowing study (VFSS) and fibreoptic endoscopic evaluation of swallowing (FEES). VFSS and FEES are complementary methods and both have advantages and disadvantages. Interventions for ND in pwMS are mainly based on functional swallowing therapy, including methods of restitution, compensation and adaptation. The aim of intervention is to prevent aspiration and aspiration pneumonia. Outcome assessment should focus on clinically relevant parameters, such as activity limitation, participation restriction and health-related quality of life.

KEY WORDS:

DYSPHAGIA; SWALLOWING; MULTIPLE SCLEROSIS

'To this symptom (dysarthria) may successively be added, especially in advanced stages of the disease, certain disorders of deglutition'

Jean-Martin Charcot, 1877

Introduction

Dysphagia is rarely an isolated, predominant symptom in multiple sclerosis (MS), but recent studies show that swallowing symptoms are much more frequent than previously assumed. For instance, in *McAlpine´s Multiple Sclerosis* the frequency of dysphagia in persons with MS (pwMS) was stated as 3% in Great Britain and 23% in Japan.[1] These figures are much lower than the real prevalence of 30–40%. Furthermore, dysphagia is life threatening, as is evident in the fact that aspiration pneumonia due to dysphagia is the leading cause of death in pwMS.[2] Accurate diagnosis and management of dysphagia may therefore help pwMS by reducing activity limitation and participation restriction and, in certain cases, by preventing death. For severely-disabled pwMS dependent on tube feeding (and in rare cases a tracheostomy), there is a therapeutic overlap between functional swallowing therapy and palliative medicine.

This article aims to give an overview of dysphagia in pwMS with special emphasis on evidence-based diagnostic and therapeutic approaches.

It has to be emphasized that the results differ between some studies mentioned in this paper, especially with regard to the frequency and methods of detecting dysphagia in pwMS. The main reason for the variation is different diagnostic methods, which range from self-reporting or clinical evaluation to instrumental methods, such as videofluoroscopy. It is problematic to rely on self reports because: silent aspirations cannot be realized by the patients themselves; and in some pwMS (especially those with advanced disease) there is often no realistic insight into swallowing disorders or cognitive dysfunction. This emphasizes the importance of accurate history taking, clinical evaluation and use of screening instruments and instrumental methods.

Definitions

Swallowing is a semi-automatic motor action of the respiratory, oropharyngeal and gastrointestinal tract muscles. It serves the dual function of transporting

ingested material from the oral cavity to the stomach, and protecting the airway from inappropriate substances.[3,4] Fifty paired striated oropharyngeal muscles and the oesophagus (containing striated as well as smooth muscles) are involved in swallowing.

Dysphagia is a disturbance of the complex sensorimotor functions of swallowing.

Neurogenic dysphagia (ND) is dysphagia resulting from a neurological disease. In ND, disturbances of the oral and/or pharyngeal phase are very frequent, in contrast to rarely-occurring oesophageal problems ('oropharyngeal dysphagia').

Other important definitions are listed in Table 1.

Epidemiology

Following a MEDLINE search in 1994[5] only one study dealing with the frequency of ND in pwMS was identified. In it, the authors state: 'Dysphagia is not a frequent complaint, but when it occurs it tends to be associated with more severe disease and is possibly lethal.'[6]

In a study on a representative group of 525 pwMS (Expanded Disability Status Scale [EDSS] scores ranging from 0 to 9.5), ND symptoms were found in 43%.[7] Comparison of the dysphagic and asymptomatic groups showed that symptomatic subjects had significantly higher EDSS scores and were significantly more impaired with regard to cerebellar, brainstem and cognitive functions. About 17% of pwMS with low disability (EDSS score 0–2.5) had ND.

A quantitative water test used in 79 consecutively admitted pwMS revealed abnormal swallowing in 43%, almost half of whom had no swallowing complaints.[8] ND was associated with abnormal brainstem/cerebellar functions, overall disability, depressed mood and low vital capacity.

An Italian study found ND in 49 out of 143 pwMS (34%), and identified a close association between ND and brainstem impairment and severity of illness.[9] This was confirmed by the De Pauw *et al.*[10] study that asked 308 consecutive pwMS whether they ever had swallowing problems: 73 had permanent ND (24%) and another 5% had a history of transitory swallowing problems. Permanent ND started to be a problem in mildly impaired patients (EDSS score 2–3) and its prevalence increased with increasing disability, to reach 65% in the most severely disabled subjects (EDSS score 8–9). Two symptoms, coughing or choking during a meal and a history of pneumonia, were present in 59% and 12%, respectively, of patients with swallowing problems. Manofluoroscopy, a combination of videofluoroscopy and manometry, showed oral phase deficiency in all 30 patients examined using this technique, but abnormalities of the pharyngeal phase were seen only in patients with an EDSS score >7.5.

Abraham and Yun[11] recently investigated 13 pwMS with ND (EDSS score ranging from 2 to 9) using videofluoroscopy. Eleven patients had primary pharyngeal dysphagia, one had primary laryngeal dysphagia and one patient had primary oral dysphagia. Upper oesophageal sphincter (UES) dysfunction was found in all pwMS in the study.

Table 1: Important definitions associated with swallowing

Pathological finding	Definition
Absent/delayed swallowing reflex	Material (food, liquid, secretions) passes the base of the tongue but does not trigger pharyngeal swallow (absent if no pharyngeal swallow is seen after three tests)
Leaking	Passage of material into the pharynx before the swallowing reflex is triggered
Retentions	Residues of material in the valleculae epiglotticae, along the pharyngeal wall or in the sinus piriformes of the hypopharynx
Penetration	Passage of material into the larynx above the level of the vocal cords
Aspiration	Passage of material into the larynx below the level of the vocal cords
Silent aspiration	Patient does not respond to an aspiration event with a spontaneous cough (most frequently caused by impaired laryngeal sensation)
Important sequelae of dysphagia	Dehydration/malnutrition, aspiration pneumonia, partial or total dependence on tube feeding (and even tracheostomy tube), decreased quality of life, increased healthcare costs, mortality

Summarizing the literature since 1994, the following statements can be made: the prevalence of ND in pwMS is high (more than 30%). ND is associated with overall disability and brainstem signs, but about 15% of pwMS with mild disability may also suffer from ND. There is no typical ND disturbance pattern for pwMS.

Anatomical Aspects

The most important parts of the forebrain, with regard to swallowing, are the anterior insula and the so-called frontoparietal operculum, which comprises the lowest part of the sensorimotor cortex and a small region of the premotor cortex. From these cortical areas, corticobulbar connections project to the ipsilateral and contralateral brainstem nuclei that are relevant for swallowing and chewing, i.e. the trigeminal (V) and facial (VII) nerves in the pons, and the glossopharyngeal, vagal and hypoglossal nerves (IX, X and XII, respectively) in the medulla oblongata.

At the brainstem level, four central pattern generators (CPGs), two on each side of the medulla, orchestrate swallowing. The ventromedial CPGs are near the nucleus ambiguus and the dorsomedial CPGs are close to the nucleus tractus solitarii (NTS). The NTS is an important brainstem nucleus that receives messages from the oral, pharyngeal and laryngeal mucosal areas as well as from the forebrain. It is therefore able to modulate deglutition depending on characteristics of the bolus, such as size, consistency and temperature.

There is an hemispheric asymmetry with regard to the representational areas mentioned, i.e. in most people one hemisphere is dominant for swallowing (independent of handedness).[12] MS lesions cause ND when the 'dominant' side of the swallowing-relevant forebrain areas are affected (the subcortical white matter more often than the cortex itself) or when both sides are affected, especially when brainstem structures (Figure 1) are involved.

The probability of swallowing-relevant cortical/subcortical areas/connections being affected increases with a higher lesion load and the number of brainstem nuclei/CPGs involved. In pwMS who are not severely disabled but suffer from ND symptoms, some of the (few) MS lesions are probably situated in strategically-relevant regions for swallowing, such as the medullary CPGs. This hypothesis may explain the association between ND severity and overall disability and/or brainstem signs.

Key Points

- *Dysphagia in pwMS is frequent (>30%) and may be life-threatening, so early diagnosis and treatment are important*
- *Diagnosis comprises history taking, clinical examination, bedside screening examinations and, often, instrumental methods such as the videofluoroscopic swallowing study or fibreoptic evaluation of swallowing*
- *The main aim of therapy is to prevent aspiration and therefore aspiration pneumonia*
- *There are many therapeutic approaches that are aimed at three principles: restitution (of disturbed functions), compensation (through swallowing techniques/manoeuvres) and adaptation (of the patient´s environment, e.g. by dietary changes such as thickening of liquids)*
- *Pharmacotherapy may help alleviate associated problems such as hypersalivation, hiccups or reflux*
- *Outcome should be assessed by clinically relevant outcome measures, including quality of life, rather than by surrogate end-points.*

Assessment of Dysphagia

Diagnosing ND in pwMS comprises history taking, neurological evaluation, bedside screening examination (BsSE) and in certain cases, instrumental methods. The usual instrumental methods are a videofluoroscopic swallowing study (VFSS) – performed as a modified barium swallow (MBS) by a radiologist and a speech/language pathologist – and/or a transnasal fibreoptic endoscopic evaluation of swallowing (FEES) (Figure 2). Most of the procedures for assessing dysphagia/aspiration that are dealt with in the following sections were not developed or validated for pwMS. They were investigated in patients with a wide range of neurological diseases, especially stroke. It is therefore only assumed that these predictors of dysphagia or aspiration are applicable to pwMS. Worth mentioning is that, with the exception of VFSS

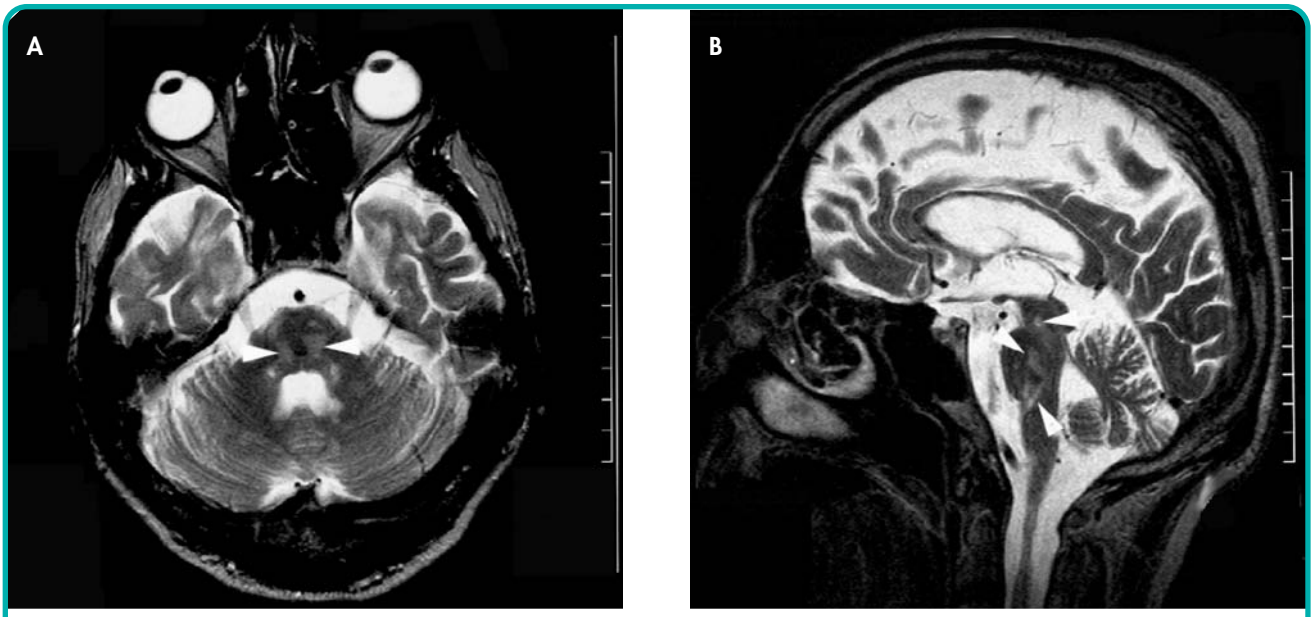

Figure 1. Magnetic resonance image (MRI) of the brain of a patient with multiple sclerosis and severe dysphagia. (A) Axial T2 image showing bilateral hyperintensities in the tegmentum of the pontomedullary junction (arrowheads pointing to only some of the lesions). (B) The sagittal T2 image shows that the hyperintensities extend from the tegmentum of the pontomedullary junction over the pons into the mesencephalon (arrowheads)

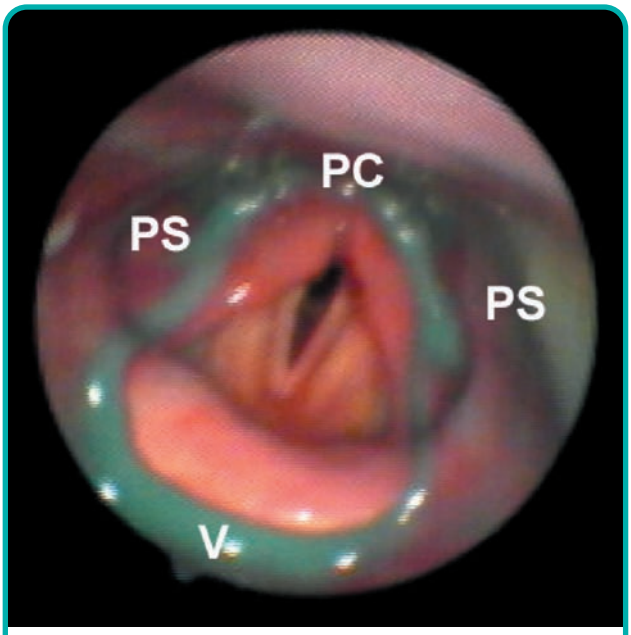

Figure 2. Transnasal fibreoptic endoscopic evaluation of swallowing (FEES) in a patient with multiple sclerosis and dysphagia. The photograph shows leaking of a liquid bolus reaching from the epiglottic valleculae (V) to the piriform sinuses (PS) and postcricoid region (PC) of the hypopharynx (without penetration into the laryngeal aditus)

and FEES, there is no single assessment instrument with a sensitivity of 80–90% and specificity of 50% or better. These values of specificity and sensitivity for VFSS and FEES have been postulated by Doggett *et al.*[13] for detecting aspiration.

History taking should include questions about weight loss, dyspnoea, cough and/or choking during or after eating or drinking, and any episodes of either unexplained fever or pneumonia. A study of 249 patients found the following clinical indicators to be good predictors of whether patients are likely to aspirate or not (correct prediction in about two thirds of patients): reclining or lying posture; dysphonia/aphonia; wet phonation; abnormal/absent laryngeal elevation; wet spontaneous cough; abnormal palatal gag on either or both sides; some or no swallowing of secretions; harsh phonation; and breathy phonation.[14] Dysphagia is necessary but not sufficient for developing aspiration pneumonia and the best predictors seem to be a dependent feeding status, dependent oral care, number of decayed teeth, tube feeding, more than one medical diagnosis, number of medications and smoking.[15]

There is insufficient evidence for guidelines regarding BsSE, but a combination of two screening tests was identified as being accurate for predicting aspiration. Patients are at risk of aspiration if they fail the 50 ml water test and have impaired pharyngeal sensation.[16] In the 50 ml water test the patient is asked to swallow 50 ml of water in 5 ml aliquots. ND is diagnosed if the patient chokes or coughs, or their voice quality alters. If any of these occur, the test is

stopped and the amount of water drunk noted. If the patient drinks all 50 ml of water without symptoms they are considered to swallow normally. Patients who perform satisfactorily on the 50 ml water test but who have respiratory or laryngeal complications of swallowing should be evaluated in more detail.

Patients with silent aspiration may seem to have no swallowing problems when performing the 50 ml water test. Reduced pharyngeal sensation correlates well with silent aspiration and so should be assessed on both sides. This is done using the tip of a cotton bud.

According to a recent study, the 50 ml water test combined with pulse oximetry may be valuable for improving the sensitivity and specificity of BsSE with regard to detecting aspiration in stroke patients. Either test should be positive to provide useful sensitivity and specificity values of 100% and 71%, respectively.[17] This result needs to be verified in larger studies and with different aetiologies.

Whether VFSS or FEES is the gold standard for diagnosing and monitoring ND is undecided, since these methods provide overlapping and complementary information.[13] The two techniques are, however, equally effective in discriminating between penetration and aspiration. According to Colodny,[18] FEES is more reliable than VFSS at assessing penetration, whereas VFSS seems to be superior to FEES in distinguishing the various categories of aspiration.

The VFSS provides an image throughout the swallow and allows viewing of the complete oropharyngeal aerodigestive tract, including the UES, which is often dysfunctional in ND. Disadvantages of VFSS are that it requires some patient cooperation and repeated use is limited (due to the radiation exposure).

Fibreoptic endoscopic evaluation of swallowing, which allows direct visualization of the pharynx and larynx before and after swallowing, may be repeated as often as necessary. It is generally well tolerated by patients, who can be tested on various food consistencies during the examination. FEES is more portable than VFSS and can be performed in bedridden patients or those unable to cooperate,[19] but does not show the morphology during swallowing itself ('white out').

The eight-point penetration–aspiration scale (PAS; 1=material does not enter the airway; 8=material enters the airway, passes below the vocal folds, and no effort is made to eject) is widely used for semi-quantitatively assessing the degree of endoscopically and radiologically measured penetration/aspiration.[20] In Germany, a four-point scale (1=penetration; 4=aspiration of >10% of the bolus volume and absent cough reflex) is used for radiologically assessing penetration/aspiration.[21]

Management of Dysphagia

The main swallowing disturbances are reduced lingual control, impaired tongue base retraction, delayed/absent pharyngeal swallow, reduced pharyngeal contraction, UES (cricopharyngeal) dysfunction, reduced laryngeal closure and diminished pharyngeal and/or laryngeal sensation. These may be found alone or in combination in pwMS.

The methods of functional swallowing therapy described focus on the dysphagic symptoms and corresponding pathophysiology, not on the stage of the underlying disease. Therapy therefore has to be tailored to the disturbed function(s) in every pwMS. The most frequent swallowing disturbances in pwMS and corresponding therapeutic methods are listed in Table 2.

Pharmacological Therapy[22]

In pwMS with severe hypersalivation, anticholinergic drugs (e.g. transdermal scopolamine lasting 72 h) or drugs with anticholinergic side-effects (e.g. amitriptyline) are effective. Botulinum toxin injection or radiation of the parotid gland is rarely indicated. When thick secretions are a major problem for the patient, N-acetylcysteine is the drug of choice. The preferred therapy for hiccup is a combination of baclofen, domperidone and a proton pump inhibitor (e.g. omeprazole), and gabapentin may be added in severe cases.

Gastroesophageal reflux disease (GERD) causes symptoms such as acid regurgitation, heartburn and/or cough, may aggravate ND and should be treated with proton pump inhibitors (e.g., pantoprazole or omeprazole). In pwMS with predominant UES dysfunction, botulinum toxin injection of the UES (endoscopically or transcervically) may be a successful intervention, but is rarely indicated. The prerequisites are the same as for cricopharyngeal myotomy, namely UES

Table 2: Frequent swallowing disturbances in persons with multiple sclerosis and the corresponding therapeutic methods of functional swallowing therapy

Disturbance	Restitution	Compensation	Adaptation
Reduced lingual control	Tongue exercises	Head anteflexion	Thickening of liquids
Impaired tongue base retraction	Tongue exercises, Masako manoeuvre	Head anteflexion, Mendelsohn manoeuvre	Smooth consistency, e.g. milk
Delayed/absent swallowing reflex	Stimulation of the faucial pillars, tongue exercises	Supraglottic swallowing, head anteflexion	Emphasizing taste or temperature of food, sour bolus
Reduced laryngeal closure	Exercises using pitch, positional, compression and respiratory support strategies, phonatory exercises	Supraglottic swallowing, turning the head to the stronger side	Thickening of liquids
Dysfunction of the upper oesophageal sphincter	Exercises for maximizing extent and timing of hyoid/laryngeal elevation, Shaker manoeuvre	Mendelsohn manoeuvre	Thin consistency
Reduced pharyngeal contraction	Whistling, sucking, snarling	Turning the head to the affected side, tilting the head to the stronger side, effortful swallowing	Smooth consistency
Diminished pharyngeal and/or laryngeal sensation	No evidence-based restitution method	Supraglottic swallowing (in the case of silent aspiration), swallowing more than once	Enhancing gustatory and thermal stimuli

dysfunction, normal elevation of the hyoid and larynx, swallowing therapy not successful in opening the UES and pharyngeal pressure sufficient to propel a bolus through the open sphincter. Manofluoroscopy is therefore necessary before the procedure.

Functional Swallowing Therapy[23]

Functional swallowing therapy can be divided into methods of restitution, compensation and adaptation.

Restitution

Restitution focuses on partial or complete restitution of disturbed functions. Effortful swallowing is indicated for pwMS with an impaired tongue base retraction and/or reduced pharyngeal propulsion. Stimulating the anterior faucial pillars effectively triggers the swallowing reflex and the combination of mechanical, thermal and gustatory stimuli seems to be more efficient than thermal stimulation alone.[24] Exercises, comprising repetitive training of sensorimotor actions, focusing on lip closure, cheek tonization, mastication, velar movement and movements of the tongue and larynx can also help. In pwMS with weakness of the suprahyoidal muscles and concomitant UES dysfunction, the Shaker manoeuvre (a repetitive head-raising exercise) may be of value. This is performed while lying in bed and comprises repeatedly raising and holding the head above the lying level.[25] The Masako manoeuvre (tongue-holding) is indicated when the approximation between the base of the tongue (BOT) and the posterior pharyngeal wall (PPW) is inadequate. The patient should swallow with the tongue stabilized anteriorly between the teeth. This leads to a better BOT to PPW approximation. Since there is an increased risk of aspiration with a bolus during this manoeuvre, it should only be performed with dry swallows.[26]

Symptoms like hypersalivation, thickening of oral secretions and sudden bouts of coughing are often caused by a reduced swallowing frequency. In those cases, the patient should be encouraged to swallow more often. Patients with MS who suffer from drooling should be advised to swallow before trying to open their mouth or speak.

Dysarthria is often associated with dysphagia, especially in pwMS with brainstem affection. Disturbances of breathing during speech and volitional control of respiration are frequent in patients with dysarthria. Exercises for improving these breathing functions are therefore of special importance.

Compensation

Compensation comprises postural changes and swallowing techniques/manoeuvres. The pwMS should sit in a comfortable, usually upright position while eating and drinking. In pwMS who have difficulty triggering the swallowing reflex, tilting the head forward may avoid leaking and subsequent aspiration. Using this 'chin tuck' widens the epiglottic valleculae and supports the epiglottic tilt. In pwMS with unilateral paresis of the tongue, pharynx and larynx, tilting the head to the stronger side may guide the bolus in this direction. Turning the head to the affected side helps close the ipsilateral recessus piriformes and prevents retentions in patients with unilateral paresis of the pharynx. When tongue movements are impaired (resulting in difficulty initiating a swallow) but the pharyngeal phase of swallowing is intact, tilting the head backwards helps guide the bolus into the pharynx.

The Mendelsohn manoeuvre is a technique that helps open the UES and prolong its opening time. The patient has to hold the upward movement of the larynx during swallowing for some seconds. This technique is appropriate for patients with pharyngeal residues or deficient opening of the UES (e.g. due to reduced laryngeal movement or weak tongue base movement). Supraglottic swallowing helps close the vocal cords during swallowing, and involves the patient holding their breath while swallowing and exhaling at full force immediately afterwards. Food or secretion can be expelled from the laryngeal vestibulum by this technique to avoid aspiration. Supraglottic swallowing is recommended when laryngeal closure is weak and/or there is a delay in triggering the swallowing reflex. It is also appropriate for pwMS who have normal or near-normal respiratory function (are able to cough and clear the throat). In cases with disturbed respiratory function, supraglottic swallowing may be impossible. It could therefore be helpful to perform respiratory exercises before supraglottic swallowing.

Adaptation

Adaptation means modifying the environment to ease nutrition. Dietary modification may help prevent extremely long mealtimes, fatigue and dread of meals. Soft textures or puréed food can compensate for a poor oral preparation phase, and ease oral and pharyngeal transport. Liquids should be thickened if thin drinks cause choking. Triggering the swallowing reflex can be enhanced by emphasizing taste or temperature; cooled drinks are often easier to swallow. An example of adaptive equipment is the nose cutout cup, which enables the patient to drink with the head tilted forward.

Safety Strategies

It is helpful to create a silent, relaxed atmosphere during mealtimes. When oral nutrition becomes difficult and needs the patient's full attention, distractions such as conversation, TV, radio and stress-inducing situations should be avoided. Patients with a significant level of fatigue are advised to eat several small calorie enriched (e.g. with maltodextrose) meals a day. Carers and families of patients who suffer from episodes of choking while eating or drinking are advised to learn how to apply the Heimlich manoeuvre as this may reassure the patient.

Tube Feeding and Tracheotomy

As ND becomes more severe, nasogastric tube feeding (NTF) and percutaneous endoscopic gastrostomy (PEG) have to be discussed. NTF should only be used for a short time because of its many disadvantages and PEG is, therefore, in most cases the preferred treatment.

Tube feeding is only indicated in pwMS who cannot eat and/or drink enough, i.e. in persons who are threatened by weight loss and/or dehydration. It has to be emphasized that tube feeding does not prevent aspiration or aspiration pneumonia. The PEG technique is not without risks, but mortality is less than 1%. The most common minor complications (about 20%) are local pain or skin infections, while the major complications (1–3%) mainly comprise peritonitis and pneumonia. Based on experiences with patients suffering from amyotrophic lateral sclerosis, the procedure should be performed as long as the vital capacity is over 50% (to reduce the risks). In pwMS with increased risk of complications during PEG insertion, due to a reduced respiratory reserve, a radiologically-inserted gastrostomy (RIG) that does not require sedation should be considered. About 4 h after PEG insertion, feeding can be initiated with

about 500 ml of tea. We begin feeding on the day after PEG insertion in the following way, which may differ from patient to patient: Day 1, 500 ml (50 ml/h); Day 2, 1000 ml (100 ml/h); Day 3, 1500 ml (150 ml/h) via a pump device. In pwMS without special problems, such as reflux or diarrhoea, the most frequently used feeding method after these first 3 days is by gravity (without a pump). Usually, we recommend that patients are not fed for a period of 8 h at night. Continuous administration via a pump is mandatory, however, for patients with a jejunostomy. As a rule, 30–40 ml of water and 25–50 kcal/kg of body weight should be administered daily. In standard feeding preparations the amount of water is 80%, i.e. 500 ml contains 500 kcal and 400 ml of water (for details see Prosiegel *et al.*[22]).

In pwMS who cannot swallow their own secretions safely, the decision as to whether or not to perform a tracheotomy has to be made. This procedure is rarely indicated. It should be considered when pwMS are threatened by choking and/or have suffered from more than one episode of aspiration pneumonia due to extreme accumulation of saliva that cannot be sufficiently removed by regular suction. The consequences of a tracheostomy should be carefully discussed with the patient, as there are several management considerations: tracheostomies have to be suctioned regularly; the tubes have to be changed; although tracheostomy itself does not cause swallowing problems in a non-dysphagic patient, it might exert a negative influence on a pre-existing dysphagia; and the tracheostomy cannula hinders speaking, since the cuff has to be blocked in the case of severe dysphagia (for details see Prosiegel *et al.*[22]).

Assessment of Outcome

Outcome measurements in pwMS and ND should be based on clinically-relevant end-points, such as mortality, morbidity and health-related quality of life (QoL). According to the International Classification of Functioning, Disability and Health (ICF) of the World Health Organization, activity limitation and participation restriction are of special importance. To assess these, the American Speech–Language Hearing Association (ASHA) National Outcome Measurement System (NOMS) swallowing scale, dietary levels/restrictions and cueing may be used.[27] Table 3 shows an ordinal scale reflecting the degree of activity limitation due to ND.[28]

Table 3: Scores for activation limitation due to oropharyngeal dysphagia[28]

Score	Activity limitation
0	Full oral, no limitations
1	Full oral, with compensation
2	Full oral, with consistency restriction
3	Full oral, with compensation and consistency restriction
4	Partial oral
5	Partial oral, with compensation
6	Tube feeding

Compensation generally involves postural changes and swallowing manoeuvres.

A valid and reliable dysphagia-related QoL-assessment instrument (SWAL-QOL and SWAL-CARE) has recently been published, but its sensitivity to treatment effects and natural history has yet to be documented.[29] Other important endpoints are nutritional measures, such as body mass index. Surrogate end-points, like the results of VFSS and FEES examinations, are important in clinical research but should not replace clinically relevant outcome measures.

Conclusions

Dysphagia is more prevalent in MS than previously thought (>30%) and is a major cause of death, so should be correctly diagnosed and managed. Diagnosis should comprise history taking, neurological evaluation and direct assessment of swallowing and its associated reflexes; once the diagnosis and cause have been established, a variety of therapies can be considered. Pharmacotherapies can be used to treat hypersalivation, problems caused by thick secretions and hiccup, as well as the symptoms of gastroesophageal reflux disease. Functional swallowing therapy focuses on completely or partially restoring the disturbed function (restitution), postural changes and swallowing techniques/ manoeuvres (compensation), and changing the

environment to ease nutrition (adaptation). Interventional therapies, such as NTF and PEG, can be considered when warranted.

Dysphagia affects quality of life, especially in terms of activity limitation and participation restriction. Outcome measures used to assess dysphagia therapy should therefore be based on clinically relevant rather than surrogate end-points.

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