

Diagnosis and treatment of bulbar symptoms in amyotrophic lateral sclerosis

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SUMMARY

Amyotrophic lateral sclerosis (ALS) is the most common neurodegenerative disease of the motor system. Bulbar symptoms such as dysphagia and dysarthria are frequent features of ALS and can result in reductions in life expectancy and quality of life. These dysfunctions are assessed by clinical examination and by use of instrumented methods such as fiberoptic evaluation of swallowing and videofluoroscopy. Laryngospasm, another well-known complication of ALS, commonly comes to light during intubation and extubation procedures in patients undergoing surgery. Laryngeal and pharyngeal complications are treated by use of an array of measures, including body positioning, compensatory techniques, voice and breathing exercises, communication devices, dietary modifications, various safety strategies, and neuropsychological assistance. Meticulous monitoring of clinical symptoms and close cooperation within a multidisciplinary team (physicians, speech and language therapists, occupational therapists, dietitians, caregivers, the patients and their relatives) are vital.

KEYWORDS amyotrophic lateral sclerosis, dysarthria, dysphagia, laryngospasm, therapeutic options

REVIEW CRITERIA

MEDLINE was used to search for papers dating back to 1995. Papers and reviews relating to amyotrophic lateral sclerosis were searched with the following terms: "ALS", "MND", "amyotrophic lateral sclerosis", "motor neuron(e) disease" and "motorneuron disease". Papers were further selected for relevance using the following terms: "dysphagia", "deglutition", "swallowing", "management", "sialorrhoea", "aspiration", "symptom control" and "palliative care". Additional papers and book chapters from the authors' own files were included when judged appropriate.

CME

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Learning objectives

Upon completion of this activity, participants should be able to:

- 1 Describe the clinical features of amyotrophic lateral sclerosis (ALS).
- 2 Identify the most common bulbar symptoms in patients with ALS.
- 3 Review the effects of upper and lower motor neuron degeneration in ALS on voice and speech.
- 4 List communication aids for managing speech and swallowing impairments in patients with ALS.
- 5 Describe options for managing salivary flow and drooling in patients with ALS.

Competing interests

The authors, the Journal Editor H Wood and the CME questions author D Lie declared no competing interests.

INTRODUCTION

Amyotrophic lateral sclerosis (ALS), the most frequent of the motor neuron diseases, is characterized by progressive degeneration of upper and lower motor neurons in the motor cortex, brainstem and spinal cord. The clinical hallmarks of ALS are progressive limb weakness, respiratory insufficiency, spasticity, hyperreflexia, and bulbar symptoms such as dysarthria and dysphagia. Neuropsychological disturbances can also occur, although these are normally subtle and cannot be detected without comprehensive neuropsychological testing.¹⁻⁴ Bulbar symptoms at ALS onset can be observed in up to 30% of patients, and almost all patients demonstrate bulbar involvement at later stages of the disease.^{5,6} Dysphagia affects food intake with the complications of

choking, malnutrition and pulmonary aspiration. Malnutrition has been shown to be an independent risk factor for death in ALS.^{7,8}

In view of the limited treatment options for ALS, palliative care and symptom control are of major interest. Progressive bulbar symptoms are often disturbing, and they result in reductions in quality of life and life expectancy.^{9,10} In this Review, we highlight the importance of early detection, as well as appropriate treatment and management, of bulbar symptoms and complications in patients with ALS, with our emphasis on a multidisciplinary approach.

PATHOPHYSIOLOGY OF BULBAR SYMPTOMS

The upper motor neuron involvement in ALS causes supranuclear symptoms, which are also known as pseudobulbar palsy. The clinical characteristics of pseudobulbar palsy are spasticity of the bulbar muscles (jaw, face, soft palate, pharynx, larynx and tongue), emotional lability (pathological laughing and crying), and a brisk jaw jerk. Degeneration of the lower motor neurons with involvement of the cranial nerve nuclei in the medulla oblongata and pons that innervate the bulbar muscles results in a bulbar palsy with flaccid pareses, muscular atrophy, and fasciculations and/or tongue fibrillations. In addition, loss of motor neurons in the spinal cord causes muscular weakness, resulting in progressive respiratory dysfunction, effortful communication and decreased voluntary as well as reflexive coughing, all of which can aggravate the symptoms of dysphagia.^{11,12}

CLINICAL PRESENTATION

Dysarthria and dysphagia are the most common bulbar symptoms in ALS. Studies have reported dysarthria in 93%, dysphagia in 86% and tongue fasciculations in 64% of patients with ALS who have bulbar symptoms.^{13,14} The clinical assessment of bulbar symptoms requires a multidisciplinary approach, comprising history taking, evaluation of weight and BMI, respiratory function assessment and clinical examination of swallowing and speech (Box 1).

Dysarthria

The underlying pathology of dysarthria is a flaccid or spastic paresis of the musculature of the face, tongue, lips, palate, pharynx and larynx. In the early stages of ALS, patients often report mild changes in voice and speech. These

Box 1 Clinical assessment of bulbar symptoms in amyotrophic lateral sclerosis.

In patients with amyotrophic lateral sclerosis (ALS), bulbar symptoms are assessed clinically by examination of the involved anatomical structures, and of the patient's ability to speak and manage food and drinks. Patients should be assessed continually to establish therapeutic strategies to maintain communication and a sufficient and secure oral intake, and to define aspiration risks.

To assess swallowing, the examiner should palpate a dry swallow with two fingers on the thyroid cartilage to assess the degree of laryngeal elevation and difficulties with swallow initiation. A test swallow with water can demonstrate incomplete pharyngeal stripping with residual water on the vocal cords; the presence of 'moist phonation' in the speech immediately after completion of the swallow is indicative of this phenomenon. A marked fall in oxygen saturation (as measured by pulse oximetry) or increased respiration rate after oral intake suggests aspiration.^{30,33}

Facial and lateral jaw movements are usually normal in the early stages of ALS. The ability to maintain a lip seal is important for the oral retention of food and fluids and permits a positive pressure within the oral cavity to initiate swallowing and facilitate laryngeal elevation. Lip strength can be tested by asking the patient to suck on the examiner's gloved finger. The ability to take food from a utensil and to use a straw should also be assessed. The tongue should be examined for fibrillations at rest, and for rapid alternating movements and range of motion, including protrusion and lateral movement. Tongue strength can be tested by having the patient press the tongue against a finger through the cheek. From the strength, rate, coordination and range of tongue movement, the ability to collect and control a bolus within the mouth and to push the bolus effectively into the pharynx can be inferred. The presence of oral residues after a swallow is a sign of disturbed tongue function.

Intact velar function is essential to create a sufficient intraoral pressure and helps to establish a negative pressure in the pharyngeal phase of swallowing. The palate can be examined by stimulating the gag reflex with a tongue blade or by asking the patient to make a prolonged "a" sound. The pharyngeal phase cannot be directly observed, but information can be gleaned through observation of swallowing. Coughing before, during or after swallowing might indicate aspiration. It is important to stress, however, that weakness of the laryngeal closure and respiratory musculature can make patients cough insufficiently or stop coughing. Pseudobulbar palsy is typically characterized by dissociation between voluntary and reflexive actions; that is, lack of elevation of the palate during phonation but a brisk palatal reflex when the palatal arch is touched.

The laryngeal structures usually appear normal in patients with ALS, although incomplete vocal fold closure, bowing and supraglottic hyperfunction can be observed.¹⁴ Examination can show both spasticity and weakness of the vocal folds, although weakness is usually predominant in the later stages of the disease. Adduction of the vocal folds during vocalization is sometimes weak, resulting in a loss of vocal volume. Abduction is usually incomplete, and the glottic airway can be limited to as little as 1 mm. In cases of pseudobulbar involvement, hyperadduction of the vocal cords is reported, whereas in bulbar involvement a pattern of hypoadduction predominates.^{30,64}

perceptible changes include a harsh, hoarse or strained voice, a breathy speech pattern with short phrases, inappropriate pauses in speech, imprecise consonants, hypernasality, and a decreased range of pitch and loudness.^{15–19}

Through a multitude of subtle movements, the tongue acts as the principal articulator of

speech. Slurring can be one of the first symptoms of impaired tongue function in ALS, and as the disease progresses patients increasingly fail to produce accurate speech, ultimately developing anarthria. The tongue can be weak and atrophic with fasciculations, or spastic with slowed movements in all directions, depending on the underlying pathology.²⁰

Flaccid or spastic paresis of the oropharyngeal muscles results in weak speech production and problems with articulation, causing impaired intelligibility of speech. Spastic orofacial muscles are typically retracted, causing problems with lip closure, whereas flaccid paresis is often associated with drooping lips and drooling of saliva.

Weakness of the soft palate and pharyngeal musculature causes hypernasality and decreased intelligibility of speech owing to insufficient nasopharyngeal closure and reduced oral airflow. These impairments result in breathy speech, and in an inability to generate long phrases owing to inadequate breath control.²¹

The laryngeal musculature controls movement of the vocal cords and larynx. The vocal cords are open during respiration, and for vocalization they move to the midline and vibrate as the airstream passes through. Changes in pitch are achieved by extension or contraction of the vocal cords from movements of the larynx. Flaccid paresis of laryngeal muscles owing to predominant lower motor neuron degeneration produces a soft, weak, low-pitched and monotonous voice. Severe upper motor neuron impairment, by contrast, makes the voice sound harsh and strained.^{22,23} Dysarthria with a spasmodic component (focal laryngeal dystonia) has also been described.¹⁷

The pattern of speech impairment in ALS usually mixes symptoms of flaccid and spastic pareses of the bulbar muscles.¹⁶ Furthermore, a decrease in respiratory function compromises speech and voice, producing, for example, effortful, breathy and strained phrasing, or affecting prosodic features such as stress, rhythm and tone, thereby limiting expressive communication.²⁰ Early detection of impaired speech and voice is crucial for timely provision of speech and language therapy in patients with ALS.

Dysphagia

Impairment of swallowing in ALS can result from weakness or spasticity of the muscles that are innervated by trigeminal, facial, hypoglossal, glossopharyngeal or vagal nerves.²⁴ During the

disease course, all muscles of the tongue, lips, palate, jaw, pharynx, larynx and upper trunk can be affected,^{25–27} resulting in inefficient bolus transport. As the motility, strength and sometimes the coordination of the orofacial and lingual muscles decrease, difficulties in oral preparation, mastication and oral transport of food emerge, which result in oral, pharyngeal and/or laryngeal residues of food or secretions. Patients with ALS generally experience difficulties in managing dry, tough-textured or crumbly food and thin liquids, although the specific nature of the difficulties depends on the patient's individual clinical pathology. Typical symptoms are an increased eating time, unintentional weight loss, and fever of unknown origin or recurrent respiratory infections. With increasing loss of tone and strength in the muscles that control lip closure and difficulty in swallowing their saliva, patients tend to drool.^{28–30} Moreover, the inability to keep the lips closed results in increased breathing through the mouth, leading to thickening of oral secretions. In addition, weakness of the hypopharyngeal muscles causes poor pharyngeal stripping during swallowing, as well as pooling of saliva in the mouth and oropharynx.

Typical features of laryngeal involvement are the failure of the larynx to move superiorly or anteriorly during the swallowing reflex, and incomplete closure of the larynx during elevation.³⁰ In the early stages of ALS, procedures such as tactile or thermal stimulation can be used to compensate for late triggering of the swallowing reflex, as described later in this article. With increasing weakness of laryngeal adduction, however, these approaches become ineffective, thereby increasing the risk of pulmonary aspiration (that is, passage of material into the larynx below the level of the vocal cords). Sensory deficits of unknown origin in the pharynx have been shown to be frequent, and these deficits further increase the risk of aspiration.^{31,32} Penetration—which, in contrast to aspiration, is defined as passage of material into the larynx above the glottic level—or aspiration without any subsequent cough are termed silent.^{23,30,33} Reduced tongue force, delayed triggering of the swallowing reflex and weak elevation of the hyoid and larynx result in reduced opening of the upper esophageal sphincter, with retention of saliva, food and liquids in the valleculae, the piriform sinuses and the postcricoid region. In addition, opening of the pharyngoesophageal segment

can be disturbed, especially in patients with pseudobulbar palsy, owing to a hyperreflexic and hypertonic upper esophageal sphincter.³⁴ A forceful cough reflex is crucial to enable clearance of aspirated food or saliva, but in patients with ALS coughing is often impaired by a progressive weakness of the respiratory and laryngeal muscles. In some cases, therefore, it will be necessary to discuss whether a patient could benefit from a tracheostomy.²³

Laryngospasm

Laryngospasm is defined clinically as a paroxysmal episode with the sensation that air cannot be moved in and out, accompanied by inspiratory stridor. This condition is caused by rapid and forceful contraction of the laryngeal sphincter, which sometimes results in complete upper airway occlusion.³⁵ Laryngospasm usually comes to light during intubation or extubation procedures in patients undergoing surgery. In one study by Sperfeld *et al.*, laryngospasm was observed in up to 50% of patients with X-linked spinobulbar muscular atrophy (Kennedy disease), whereas only 2% of the control group, who were in the early stages of ALS, reported this symptom.³⁶ Laryngospasm was reported in about 19% of another group of patients who were in the later stages of ALS.³⁷ Smoking and gastroesophageal reflux seem to be important risk factors for laryngospasm. On the basis of gastroscopy findings, Sperfeld *et al.* diagnosed gastroesophageal reflux in 3 out of 15 (20%) patients who had Kennedy disease and laryngospasm.³⁶ It is currently assumed that laryngospasm is caused by a combination of gastroesophageal reflux, aspiration of gastric contents, and functional impairment of neurons involved in swallowing and regurgitation.

THERAPY FOR SPEECH IMPAIRMENT

Various general strategies, as well as more-specialized speech and language therapies, can be employed to facilitate speech in patients with ALS. Positioning and physical comfort can decrease abnormal muscle tone and prevent or reduce reflexive responses or spasticity. Furthermore, an optimized body position reduces the effort required to maintain body position and respiration. Physiotherapists or occupational therapists can be very helpful in this regard. Background noise, such as that from a television or radio, and group settings with many people talking at the same time, should be avoided.²⁰

Compensatory speech techniques

If disease progression is slow, speech therapy can help to correct ineffective compensation strategies in patients with ALS. In the early stages of the disease, for example, patients tend to use too much force for their voice and articulation as they become aware of difficulties in being understood. This forceful speech, as well as an increasing loss of breath, often leads to a further deterioration of intelligibility. Patients can learn and perform breathing and relaxation exercises to optimize usage of the available respiratory resources. In speech and language therapy, patients are instructed to use their voice and articulation patterns, as well as their breathing rate during speech, in the most economical way, so as to optimize usage of the available respiratory resources (Lindner-Pfleghar B *et al.*, unpublished data). In addition, facilitation techniques, such as vibration, or application of ice to the involved musculature, can help to decrease muscle tone and improve speech intelligibility for a short time period.^{20,38}

In patients with ALS who have rapidly progressing disease, voice and articulation therapies do not seem to produce notable therapeutic effects; in fact, resistance and isometric exercises, oral motility, voice strengthening training and loudness practices can cause a decrease in voice quality and intelligibility.¹⁹

Communication aids

As dysarthria progresses, the attending speech and language therapist or physician must initiate timely provision of augmentative and alternative communication devices. Beukelman and Ball revealed a rapid decline in sentence intelligibility when the speaking rate dropped below 100 words per minute in patients with acquired neurogenic disorders.³⁹

The most simple and economical communication aids are pencil and paper, an alphabet board, and word or picture boards. The patient must, however, have sufficient function of the upper extremities in order to use these aids. If such tools are no longer an option, other augmentative communication devices are necessary. For example, laser pointers fixed on glasses or a headband can be used in combination with an alphabet board. Electronic communication devices with a keyboard or a scanner to detect head or eye movements and with a voice output enable patients to use telephones and computers in a very effective way.^{20,40} Patients can be sent

for recording of their voice patterns before they develop severe dysarthria so that the communicative device can be programmed with their own voice instead of a robotic sound. Timely education about future prospects helps the patient to accept the situation, and early adaptation to a communication device makes it easier for the patient to learn how to use it.

THERAPY FOR SWALLOWING IMPAIRMENT

To facilitate swallowing, patients with ALS should be brought into a normal physiological position while eating and drinking—usually upright, and supported by adjuvants (e.g. cushions and specially designed wheelchairs, tables, spoons and cups) if necessary. Although marked improvement in swallowing is an unrealistic goal in this progressive disease, compensatory and restitutive swallowing therapeutic methods can help to ensure oral alimentation for as long as possible and also help prevent food aspiration.²³ An occupational therapist can help by providing instruction on the optimum use of remaining body functions, and by introducing special aids, such as utensils that are easier to grip for patients with impaired hand function. The patient or their carers should be advised to practice meticulous hygiene of the oral cavity, as persisting oral residues could cause bacterial infections of the airways.

Compensatory methods

Swallowing in patients with ALS can be improved by changes of posture, or by the use of special swallowing techniques. The various methods should be used in modified form, taking into account the special disease characteristics. It is important to train not only the patient, but also the carer, as the carer can help the patient to use methods of compensation during swallowing, as well as prepare appropriate kinds of meal (e.g. fluid, thickened or solid).

Patients with tongue weakness or reduced tongue mobility can tilt their heads backwards to support the transport of the food bolus by gravity. Problems with mastication can necessitate a change of diet to a fluid or moist consistency. If the problem is associated with a delayed triggering of the pharyngeal swallow or leaking of food into the pharynx, chin tuck (tilting the chin down before swallowing) is a helpful compensatory change of posture. This position widens the vallecular space, helps to divert food away from the laryngeal vestibule,

and shortens the passage along which food is transported, thereby reducing the risk of aspiration.⁴¹ To avoid drooling of food or liquids, patients should be encouraged to seal their lips with their hand.

In cases in which predeglutitive or intrade-glutitive aspiration caused by weak laryngeal closure or a delayed swallowing reflex is diagnosed (Box 2), the supraglottic swallowing maneuver helps to close the vocal cords during swallowing (glottal closure). In this maneuver, patients hold their breath while swallowing and exhale at full force immediately afterwards.⁴² This method enables food or secretions to be expelled from the laryngeal vestibule in order to avoid aspiration. The technique is appropriate for patients with minor oral, laryngeal and/or respiratory dysfunction.

Restitutive methods

Sensorimotor exercises aimed at partial restitution of impaired functions might provide some limited benefits to patients with ALS who have swallowing impairments, although intensive exercises might merely exhaust the weakened muscles.

If the triggering of the swallowing reflex is delayed or patients show impaired oral bolus control, tactile or thermal stimulation of the anterior faucial pillars and the tongue before meal times might produce a temporary improvement. Passive pressure on the hyoid bone (either by the patient or by an assisting nurse or relative) during eating can support the elevation of the tongue and thereby trigger the swallowing reflex.

Dietary modifications

During meal times, patients with ALS should avoid distractions such as conversation, television or radio. Patients who experience appreciable levels of fatigue are advised to eat their food as several small meals a day. If an episode of choking occurs, the Heimlich maneuver can be applied by the carer.

In mild or moderate dysphagia, dietary modification has been shown to be an effective approach. Dieticians can give advice on how to enrich meals by use of foods high in calories, proteins and vitamins, and by the addition of high-energy supplements.^{43,44} In cases of constipation caused by abdominal weakness and failure of glottic closure, dietary fiber can be added to the diet. The triggering of the swallowing reflex can be enhanced by emphasizing taste or

temperature; for example, drinks can be made easier to swallow by cooling them to below room temperature. Special eating or drinking aids can also be employed. In patients whose swallow is delayed, thickened fluids are usually better tolerated than thin fluids as they tend to move more slowly through the oral cavity. For these patients, it is recommended that thickeners in powder form are added to drinks to minimize the risk of penetration and aspiration.⁴⁵ Soft textures or puréed food can compensate for a poor oral preparation phase and ease oral and pharyngeal transport. Liquid supplements can be helpful, but beyond a certain stage in the disease choking can become frequent, especially with thin liquids. Clinical and instrumental investigations (Boxes 1 and 2) are required to establish whether an individual patient will benefit from fluids or thickened drinks.

In cases of slowed eating, encouraging patients to take longer over meals is not necessarily beneficial; for instance, slow eating can induce earlier satiety, or some patients may be embarrassed that they are taking longer than others. In cases of very slowed eating, therefore, a 'little and often' approach to meals should be encouraged.⁴⁵

Augmented feeding techniques

As dysphagia increases and oral food intake becomes impossible because of exceedingly prolonged meal times or frequent choking, enteral nutrition should be considered. Careful discussion with the patient and the patient's family is required to ensure timely provision of enteral nutrition, for reasons that we will discuss below.

Fine-bore nasogastric tubes can be used for short-term feeding, although they can easily become displaced and are very visible and uncomfortable for the patient. Furthermore, these tubes carry a high risk of ulceration and aspiration pneumonia, as well as of oropharyngeal secretions.⁴⁶ For medium-term and long-term purposes, enteral nutrition should be administered via percutaneous endoscopic gastrostomy (PEG) or percutaneous endoscopic jejunostomy (PEJ).⁴⁷ In patients who have a marked respiratory impairment (vital capacity <50%), a radiologically inserted gastrostomy (RIG) can be considered. The advantage of this last approach is that it does not generally require sedation during insertion,⁴⁸ and it might, therefore, be safer than PEG under these circumstances.⁴⁹

Box 2 Instrumental assessment of bulbar symptoms in amyotrophic lateral sclerosis.

Videofluoroscopic study of swallowing (VFSS) is an important technique in the evaluation of swallowing disorders in patients with amyotrophic lateral sclerosis, as it provides an image of the entire duration of the swallow and enables the complete oropharyngeal tract, including the upper esophageal sphincter and the esophagus, to be viewed.²³ On the basis of VFSS observations, a treatment plan for swallowing therapy can be established and a decision made as to whether to provide dietary modification, or whether to instigate alternative feeding methods or compensatory positioning of the head.^{65,66}

Some clinicians advocate the use of a modified barium swallow to test for aspiration pneumonia in patients with ALS, although great care should be taken owing to the frequent finding of respiratory dysfunction and the high risk of aspiration in patients with dysphagia.⁶⁷ Every precaution should be taken to avoid barium aspiration, which is likely to cause stress to the patient. The isosmolar contrast agent Iotrolan, which has no significant adverse effects even in the case of aspiration, is recommended as an alternative.⁶⁸ The penetration–aspiration scale (PAS) is used to judge the degree of aspiration.⁶⁹

Like VFSS, transnasal fiberoptic endoscopic evaluation of swallowing (FEES) enables direct observation of predeglutitive and postdeglutitive actions within the pharynx and larynx. No observation is possible during the intradeglutitive phase as muscle contraction in the larynx and pharynx brings the mucous membrane into contact with the endoscope.⁷⁰ FEES is generally well tolerated by patients, and real food can be eaten during the examination. Complications such as epistaxis or vasovagal syncopes are rare, and FEES is more portable than VFSS and can even be performed in bedridden patients.

In conclusion, VFSS and FEES are complementary methods and both have advantages and disadvantages.⁷¹ The techniques are useful for educating patients and carers, as they clearly reveal the process of swallowing and demonstrate the risks involved in swallowing and the benefits of modifications to the feeding process.⁷²

It is important that the option of enteral nutrition is offered early in the disease, as the risk associated with the insertion of a PEG or PEJ tube is relatively low at this stage. Enteral feeding can initially be used as a supplement to ongoing oral intake. To minimize the risks of the procedure, it has been suggested that the issue of

enteral nutrition should be raised with the patient soon after the onset of dysphagia, when the patient's forced vital capacity is still over 50% of the predicted value.^{50,51} Although major complications such as peritonitis, necrotizing fasciitis, hemorrhage and death have been described, PEG or PEJ tube insertion is usually safe, and only mild complications such as local skin infections, reflux, gastric ulcer, or mechanical problems with the tube are usually observed.⁵² A study of 35 patients with ALS demonstrated an increasing BMI after insertion of a PEG tube, as well as a lower mortality rate in comparison with patients with ALS who did not receive a PEG.⁵³ Other studies have indicated improvements in quality of life, as well as maintenance of or a gain in weight, after introduction of enteral nutrition in patients with ALS.⁵⁴

Pharmacological interventions

In patients with ALS who have drooling problems, it is possible to reduce salivary flow with transdermal patches that dispense scopolamine (hyoscine hydrobromide) over a period of 72 hours,⁵⁵ or with atropine or drugs with anticholinergic effects, such as amitriptyline or doxepin.⁴⁴ Botulinum toxin injection into the parotid glands has been shown to be an efficient and cost-effective alternative to these approaches.⁵⁶ Local radiation of the salivary glands with 7–7.5 Gy is an experimental therapy for drooling that is rarely used at present.⁵⁷ Many patients worry about choking to death and find it reassuring to have a suction machine to hand. Another major problem for patients who drool is the lip excoriation that occurs with the continuous rubbing of the mouth. Patients can be instructed to dab away the saliva instead of rubbing the mouth, and to use petroleum jelly or other lip protectors.

Thick mucous secretions are frequently reported by patients with ALS and can be a sign of dehydration. Dehydration can occur because patients are unable to swallow fluids or are drooling large amounts of saliva that would usually be swallowed and reabsorbed. Hydration is, therefore, an important first step in the treatment of these patients. Liquefaction of thick mucous secretions by use of *N*-acetylcysteine might also be helpful. Manual support by a physiotherapist and mechanical cough assisting devices (insufflators) are vital for patients who have thick mucous secretions combined with a weak cough pressure.^{58,59}

Surgical options

Palliative surgery to treat dysphagia in patients with ALS should be considered only in selected cases, and even then with caution, as surgical intervention and general anesthesia might contribute to neuronal death and a more progressive disease course.^{60,61} The efficacy of cricopharyngeal myotomy in patients with ALS who have disturbed opening of the upper esophageal sphincter remains controversial (the same holds true for botulinum toxin injection into the sphincter). In patients who are unable to swallow their own secretions safely and therefore experience severe aspiration of saliva (perhaps with subsequent pneumonia), a tracheostomy with insertion of a blocked tracheal cannula might be necessary.⁶² It is important, however, to discuss this procedure and its consequences (i.e. life-prolonging effect with ongoing worsening of motor function) with patients and their relatives in detail before going ahead.

THERAPY FOR LARYNGOSPASM

Rapid upright positioning of the trunk or fixation of the arms to stabilize the body were found to be sufficient maneuvers to shorten episodes of laryngospasm in patients with ALS.³⁶ Long-term therapy for laryngospasm should consist primarily of lifestyle modifications, including appropriate spacing of meals and avoidance of late-night meals. In addition, medications that increase gastric acid secretion should be avoided. In general, avoidance of alterations of the upper aerodigestive tract, and gastroesophageal reflux treatment with prokinetic and antisecretory drugs, can reduce the frequency of attacks.^{36,63}

CONCLUSIONS

Optimal care of patients with ALS requires attention to laryngeal and pharyngeal symptoms. With the assistance of a speech and language therapist, a physiotherapist, an occupational therapist and a dietitian, the patient can overcome mild to moderate degrees of limited activity. The adoption of special techniques can aid swallowing and communication. In advanced dysphagia, augmented feeding strategies should be considered, and progressive dysarthria requires the use of special communication devices. In summary, comprehensive clinical care of patients with ALS requires a multidisciplinary team to increase life expectancy and enhance quality of life.

KEY POINTS

- Bulbar symptoms such as dysarthria and dysphagia are frequent features of amyotrophic lateral sclerosis (ALS) and can reduce life expectancy and quality of life
- Dysarthria results from flaccid or spastic paresis of the musculature of the face, tongue, lips, palate, pharynx and larynx
- Dysphagia can result from weakness or spasticity of the muscles innervated by trigeminal, facial, hypoglossal, glossopharyngeal or vagal nerves
- Laryngospasm, which is defined as a paroxysmal episode with the sensation that air cannot be moved in and out, accompanied by inspiratory stridor, affects up to 19% of patients with ALS
- Speech therapy can be helpful in the early stages of ALS to correct ineffective compensation strategies; augmentative and alternative communication devices should be provided as the disease progresses
- Compensatory and restitutive methods and dietary modifications should be used to ensure oral alimentation for as long as possible and to prevent aspiration of food; enteral nutrition should be considered as dysphagia increases

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