A Swallowing & Speech Exam for Patients with Motor Neuron Disease

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What Information Is Obtained in a Clinical Examination of Speech and Swallowing?
The following examination is carried out during a patient’s initial clinic visit, and an abbreviated form is conducted on follow-up visits. We have organized the evaluation into three general areas:

1. **History**, in which information is obtained about the nature and course of the disorder, as well as about how the disorder is being managed
2. **Physical examination**, in which the physiologic abnormalities in the speech and swallowing mechanism are described
3. **Assessment of speech and swallowing function**, in which impact of the impairment is assessed with regard to the patient’s ability to communicate efficiently and understandably, to maintain adequate nutrition and hydration, and to handle oral secretions adequately

The “Clinical Examination for ALS” form in Chapter Appendix 1.1 summarizes the information obtained during the clinical examination. During the course of the clinical examination, be cautious not to attribute all of the individual’s problems to the neurologic disorder. Unfortunately, degenerative neurologic diseases are not an antidote to other disorders or conditions that might be present.

The History
Taking the patient’s history sets the stage for the remainder of the clinical examination. We are interested in the nature of the illness, its length, and specific details concerning its onset and diagnosis. We are also interested in how patients and their families are handling the speech and swallowing problems.

Patient Information
The standard patient information contained in the Clinical Examination form is obtained. Patient information includes not only demographic data, but also the referral source. It is important to know from whom the referral came. In the case of our clinic, referrals come primarily from neurologists, physiatrists, specialty clinics such as the muscular dystrophy clinic, and community agencies such as Home Health Care. We seek to know why the referral was made from two perspectives—that of the referring health-care specialist and that of the patient.

Medical History
In obtaining the past medical history, we are interested in the neurologic disease, when it was diagnosed, and by whom. With many diseases such as ALS, diagnosis is at times complex. Therefore, if the individual has not sought a second opinion, we encourage doing so. Again, because of the complexity of the diagnosis, it is not unusual for the diagnosis to be made months and even years.
after the onset of first symptoms. Therefore, we want to know the approximate date of onset of the symptoms. What were the first symptoms? When did they begin? How have they progressed? We take particularly detailed information about the onset and progression of the speech and swallowing symptoms. Finally, when discussing the symptoms associated with degenerative neurologic diseases, it is important to obtain an understanding of the patient’s level of knowledge of the disease process. This can range widely from individuals and families who are well read and conversant with the “statistics” of the disorder, to patients and families with little or no knowledge of the disease. The patient’s desire to understand the disease process also varies widely: Some patients wish to read all they can about the disorder, whereas others wish to know no further information about it.

When taking the history, we are interested not only in the current illness, but also in the person’s history of other disorders. Although the disease cannot be cured, care should be taken not to overlook other potentially treatable disorders (such as esophageal webs, esophageal masses, or gastroesophageal reflux) and not to wrongly attribute all symptoms to the known degenerative process.

A psychosocial history is also critical in assisting the individual with a degenerative disorder to make plans for the future. We need to know, for example, the person’s employment status and how the disease has affected it. Information related to the person’s living situation and household members is also obtained.

Current Health-Care Management
During a discussion of the patient’s current health-care management, we first review the medications that the patient is taking. Combinations of medications and their side effects may have important impacts on speech, swallowing, and saliva management. When considering saliva management, we often suggest that this is a good time to review with the primary-care physician all of the medications currently being taken and to consider the elimination of potentially unnecessary ones. With a diagnosis of ALS, the priority of health-care issues may change. Medications such as diuretics used to treat a cardiac condition may not be required, given this new life-threatening disease and may even complicate management of ALS symptoms. The review of current health-care management also allows us to evaluate how effectively the patient is using the health-care delivery system. We are interested in the following types of information:

1. What community resources is the person aware of or interested in exploring?
2. What adaptive equipment does the person currently have? When was it obtained? From whom was it obtained?
3. Does the person make use of attendant care to assist in activities of daily living?

For people who are difficult to understand because of severe dysarthria, we are interested in knowing whether an emergency communication system is in place. For those with swallowing difficulties, we are interested in knowing whether the patients and family members have been given instruction in
administering first aid for choking and whether an emergency plan in case of choking is in place.

Many individuals with ALS establish a “living will,” in which they make known their medical management wishes. These instructions regarding the level of medical care they wish to receive are implemented in cases in which patients are unable to make their wishes known to the health-care management team. Thus, it is important to know whether the patient has established a living will and the specific wishes of the person in case of emergency.

The Physical Examination

During the physical examination, the impairment or physiologic abnormalities of the speech and swallowing mechanisms are evaluated. The physical examination is organized into general groups according to components of the speech and swallowing mechanisms.

Group 1: Tongue and Lips

Lip weakness can be detected by asking the patient to suck on a gloved finger (i.e., a suckling movement) or evaluated by asking the patient to blow out the cheeks. (Note that velopharyngeal competency is also required for the latter activity.) Lip weakness is also associated with the following progression: inability to whistle, food spillage, inability to use a straw, dysarthria, and lastly, drooling.

We use diadochokinetic rates or alternate motion rates in order to obtain an “objective” measure of the impairment of the lips, anterior and posterior tongue, and mandible (Darley et al., 1975). Alternate movement rates give an indication of the speed and regularity of reciprocal muscle movements. Normative data for alternate movement rates have been reported by a number of researchers (Dworkin & Aronson, 1986; Kreul, 1972; Lass & Sandusky, 1971; Ptacek, Sander, Mahoney, & Jackson, 1966). Generally, the mean syllable repetition rates for normal adults range from 5.8 to 6.9 syllables per second for repetitions of /p/ and /t/. Repetition rates for /k/ range from 5.2 to 6.2 for adults. Table 1.11 contains data taken from Dworkin and Aronson (1986) that may be useful as a measure against which to compare the clinical population. The table contains alternate motion rates (in syllables per second) for the syllables /p/, /t/, and /k/ obtained from a group of individuals without neurologic impairment and from a group of individuals with ALS. Normal males (N = 67) had a mean age of 38.5 years, and normal females (N = 58) had a mean age of 37 years, with a range from 20 to 72 years. The individuals with ALS (11 men and 8 women) had both flaccid and spastic components and represented various degrees of speech impairment.

Alternate motion rates may be measured in a number of ways. For example, the number of syllables produced by the speaker within a specified time period can be counted, or the speaker can be asked to produce a specified number of syllables and the time for that production measured. In a population in which generalized weakness is a problem, the combination of poor respiratory support and an inefficient velopharyngeal valve may make the production of long syllable sequences impossible. Therefore, we give the following instructions when asking a patient to perform the alternate movement rate tasks:
I am going to ask you to say a series of syllables as rapidly and precisely as you can. Go as fast as you can but make each syllable as distinct as you can. Be sure to take a deep breath before you start.  

We obtain alternate movement rates for /p/, /t/, /k/, and /pataka/ and measure the time required to produce a sequence of five syllables. When the person has insufficient respiratory support to produce five syllables on one breath, we measure the time required to produce however many syllables the person can say on a single breath. Although alternative motion rates give some useful information related to tongue and lip impairment, results should be interpreted with some caution. A severely impaired speaker must compromise between the precision with which the syllable is produced and the rate of production. In other words, rate can be increased if precision is sacrificed and vice versa. As with any of the measures we obtain as part of the physical examination, alternate movement rates cannot be interpreted apart from other measures.

During the physical examination of swallowing, dysfunction of the lips is most easily seen by food spillage from the corners of the mouth. The earliest symptom of this dysfunction is an increased use of napkins during meals. Dysfunction of the lips may also be noted and described as the inability of the lips to constrict around the contour of the spoon to strip the food out of the bowl and retain it in the oral cavity. Dysfunction of the tongue is indicated by food debris sitting in the gingival sulcus of the mouth after swallowing, as well as by the need for extensive mastication. This dysfunction is due to the inability of the tongue, in conjunction with the buccal muscles, to gather the food on the lingual side of the teeth to form a bolus prior to the initiation of a swallow.

**Group 2: Palate, Pharyngeal Constrictors, and Muscles of Mastication**

Palatal function can be assessed by visual examination as well as by articulation testing. By observing a gag reflex, you can find out whether the oral pharyngeal muscles constrict and the palate elevates symmetrically and completely, elevates to Passavant’s pad but weakly, or fails to meet Passavant’s pad. Palatal impairment may result in a number of speech production problems, including the following:

1. Inability to produce accurately many of the speech sounds that require a buildup of intraoral air pressure, such as /p/, /b/, /t/, /s/, or /θ/
2. Nasalization of vowel production
3. Escape of air through the nose (nasal emission) rather than oral airflow

To test for more advanced palatal dysfunction, check for nasal air escape when the patient puffs out the cheeks. Progressive symptoms associated with palatal dysfunction are hypernasal speech, inability to use a straw, and lastly, nasal reflux. In our clinical experience, nasal reflux is not common in ALS (less than 5%) despite total palatal dysfunction in some patients. Individuals with ALS, even those with near-total palatal dysfunction, often cannot generate enough oropharyngeal constrictor pressure to cause the reflux that occurs in other patients.
with palatal dysfunction. Weakness of the soft palate may also cause ineffective transport of the food bolus from the anterior to the posterior part of the oral cavity. The palate must function in concert with the tongue in positioning the bolus and acting as a scaffold against which the tongue can propel the food bolus posteriorly. Pharyngeal constrictor weakness may be most evident in the patient’s swallowing dysfunction. Specifically, the person may have difficulties clearing leafy vegetables, such as lettuce or cabbage; liquids may penetrate the airway before a swallow is triggered; and pooling in the hypopharynx may be evident due to inadequate pharyngeal stripping.

The deterioration of the muscles of mastication is difficult to assess clinically. Palpation of the masseter and temporalis muscles during biting movements can demonstrate muscle wasting. The pterygoid muscles can be assessed grossly by asking the patient to move the jaw from side to side. Few symptoms are noted early in the development of weakness in the muscles of mastication. Usually there are parallel developments in the oral cavity and pharynx that limit other aspects of swallowing ability. These simultaneous impairments may mask decreased ability to chew. In advanced bulbar ALS, weakness of the muscles of mastication may cause the jaw to be pulled downward by gravity, which leaves the mouth open. The pattern of airflow during rest breathing is changed from nasal to oral breathing, with the resultant problems of dry lips, dry mouth, and tenacious oral secretions.

Group 3: The Buccal/Frontal/Orbital Muscles (Facial Nerve), Sternocleidomastoid/Trapezius Muscle (Accessory Nerve), and the Vocal Folds (Vagus Nerve)

While the lower branches of the facial nerve (lips) are affected early in the course of bulbar disease, the upper branches of the facial nerve become involved later, with the result that the orbital and frontal muscles are affected least and latest. The sterno-cleidomastoid and trapezium muscles vary in the degree to which they are affected and, unlike the other cranial nerves, are often asymmetrically involved. Sternocleidomastoid involvement is manifested in an inability to keep the head from falling forward, and involvement of the trapezium in an inability to raise the arms—both symptoms being found in classical 11th nerve syndrome.

Examination of the vocal folds shows that most patients are able to adduct their vocal folds and phonate, even with rather advanced disease. In our experience, however, vocal fold abduction is impaired in 30% of patients and is usually symmetrically affected. Some patients, even on their initial indirect examination, are found to have the vocal folds nearly paralyzed in the midline. This finding may not be correlated with audible stridor if the respiratory muscles are too weak to generate sufficient negative pressures and the narrowed glottis does not present a functional restriction of the airway. In advanced cases, passive paradoxical movements of the vocal folds can be noted.

Our assessment of vocal fold function includes a prolonged phonation task in which the patient is asked to take a breath and say “ah” as long, steadily, and clearly as possible. This task is a good indicator of respiratory support for speech as well as vocal fold function. We listen for the following features:
• **Voice quality.** Does the voice sound smooth and clear in quality or does it lack full tonal clarity? Two general categories of dysphonic voices have been identified in neurologic disease (Darley et al., 1975). **Breathiness** is defined as a lack of fullness of voice associated with excess air wastage. Breathiness is frequently associated with flaccid paralysis. **Hoarseness** is the term used to describe a variety of noisy voices. Among the most important voice changes in ALS is the “strained-strangled” voice quality associated with spasticity, and the “wet” or “gurgly” voice quality associated with the accumulation of secretions in the larynx.

• **Duration.** Maximum sustained phonation is related to vital capacity, pitch, loudness, the function of the vocal folds, and the degree of effort used by the speaker during each trial. Phonation time was found to be a reliable predictor of vital capacity in a group of individuals with ALS (Hillel, Yorkston, & Miller, 1989). In a study of 41 individuals with ALS, variation in sustained phonation time predicted almost 50% of the variability in vital capacity. Abnormal muscle tone of the vocal folds may also affect maximum phonation time. Generally, the greater the breathiness, the shorter the duration of phonation. Phonation times may also be shortened in severe cases of strained-strangled harshness (Darley et al., 1975).

• **Pitch and steadiness.** Although pitch and steadiness alterations in voice quality are found in many neurologic disorders, they are relatively uncommon in the ALS population.

• **Loudness.** Because of the close relationship between respiratory support and voice loudness, voices that are inadequately loud should be noted. Patients with reduced voice loudness will also frequently complain of fatigue after extended periods of speaking.

• **Ability to initiate a quiet voice.** This task is a good indication of fine control of the laryngeal musculature because it removes some of the respiratory demands involved in the production of louder phonation.

• **Alternate movement rates for phonation.** We also obtain alternate motion rates for the vowel /a/ versus no phonation. This task is particularly sensitive to problems in respiratory and phonatory control.

**Group 4: The Muscles of Respiration**

Although respiratory deterioration does not appear to have a clear pattern of progression in relation to other physical findings (Braun, 1987; Fallat & Norris, 1980), it usually occurs as bulbar/spinal symptoms progress (Griggs, Donohoe, Utll, & Goldblatt, 1980). Respiratory function can be measured clinically by assessing the depth and volume of respiration, as well as cough force and phonation time (Hillel, Yorkston, et al., 1989). However, other factors (vocal fold dysfunction and secretions) can obscure the accuracy of these measurements.

Early respiratory weakness is most sensitively measured by pulmonary function tests. A limited but simpler measurement uses a handheld respirometer to assess vital capacity (VC) (Fallat et al., 1979). Normal values for VC range from 2 to 7 liters, with the peak of the distribution curve at about 4 liters. The
“expected” VC for an individual is affected by weight, height, age, sex, race, ethnic background, and altitude (Clausen, 1982). In more advanced stages of bulbar ALS, respirometers become difficult to use, as the patient cannot make a good seal around the mouthpiece, and laryngeal abductor weakness may restrict expired airflow. The formulas for predicted vital capacity based on age and height are as follows:

**Men:** NVC \( = (–38.3 \text{ age}) + (121.3 \text{ height in inches}) + 2100 \times 5 \text{ VC} = 970 \text{ cc} \\
**Women:** NVC \( = (–22.3 \text{ age}) + (110.3 \text{ height in inches}) + 2980 \times 5 \text{ VC} = 790 \text{ cc}

**Assessment of Speech and Swallowing Function**

The final phase of the clinical examination involves the documentation of the speech and swallowing functions. In other words, how has the impairment in the speech and swallowing mechanisms affected the person’s ability to communicate understandably and efficiently, to maintain adequate fluid and nutritional intake, and to manage oral secretions in an acceptable manner? Studies suggest that there is a moderate to strong correlation between measures of speech function, as measured using intelligible words per minute, and motor function, as measured by the physical examination (Yorkston, Strand, & Hume, 1998). Factors such as cognition and rate of disease progress may contribute the lack of one-to-one relationships between these two measures of function.

**Speech Function**

A number of measures of overall speech function have been proposed (Yorkston, Beukelman, Strand, & Bell, 1999), including speech intelligibility, speaking rate, articulation adequacy, and speech naturalness. Of these measures, speech intelligibility is perhaps the most important in developing a communication management plan for an individual with degenerative neurologic disorders. In our clinic, we use sentence intelligibility along with speaking rate (Yorkston, Beukelman, & Tice, 1996) as a measure of speech function. Because fatigue is a concern, we shorten the standard test length by half, requiring the speaker to produce only one sentence at each sentence length. This results in a sample that is 110 words in length.

As the severity of the dysarthria increases, the sentence production task becomes increasingly fatiguing for the speaker, and little clinically useful information can be derived from it. Therefore, once sentence intelligibility has fallen below 30%, we begin to document the level of speech disability using a single-word production task. We developed a series of sets of semantically related words—for example, numbers from 1 to 20, colors, holidays, items of clothing, and others. The single words produced by the individual with ALS are audiotaped and judged both with and without context. We have found that single-word intelligibility within semantic context provides a useful indicator of how functional natural speech is in the severely dysarthric individual (Hammen, Yorkston, & Dowden, 1991; Yorkston, Hammen, & Dowden, 1991).

**Swallowing Function**

Perhaps the best overall measure of severity of dysphagia is obtained by comparing the current weight of the patient with previously recorded weights. The
clinical examination should include a test swallow in order to evaluate the oral pharyngeal components of swallowing and assess the appropriateness of the patient’s current swallowing technique. For example, has the person learned to tuck the chin to the chest when attempting to initiate a swallow?

Motion radiographic studies of swallowing (videofluoroscopic barium swallows) are used to evaluate swallowing in individuals with symptoms that cannot be explained on the basis of the physical examination. Because this population has previously diagnosed disease, most of the symptoms of swallowing impairment are predictable and consistent with the physical findings. In a small percentage of cases, however, when the complaints do not match the physical findings or the complaints and history suggest multiple problems—such as specific cricopharyngeal dysfunction, esophageal webs, diverticuli, esophageal dysmotility, or gastroesophageal reflux—radiographic studies are helpful. Because aspiration is evident on the basis of the physical examination and history, radiographic studies generally should not be used for the detection of aspiration alone. One exception is when the clinician is willing to recommend discontinuing oral intake and the patient is willing to accept this recommendation if airway penetration is evident. When motion radiographic swallows are indicated and ordered, the studies should include a complete assessment of all stages of swallowing to allow evaluation of oral function, pharyngeal contractions, laryngeal airway protection, esophageal motility, and gastroesophageal function, as well as the morphology of all these mechanisms.

In addition to the physical findings, detailed questions about the patient’s swallowing complaints provide useful information regarding the severity of the swallowing disability. In progressive degenerative disorders, swallowing problems often have a slow onset. Thus, it is important to identify the early warning signs of progressive disorders so they can be managed effectively in order to avert further complications. The following are a series of complaints that can be viewed as early warning signs of a swallowing problem:

- **Complaints of difficulty with speech.** Dysphagia usually parallels or shortly follows the development of speech problems. If a patient is complaining of speech difficulty but not a swallowing problem, question him or her carefully to detect the possibility of an early swallowing disorder.
- **Decrease of food intake or alteration in diet.** Weight loss is often wrongly attributed to other symptoms (such as depression, malaise, or loss of muscle mass) but is often due to dysphagia.
- **Food spillage from the oral cavity.** Food spillage from the oral cavity results from discoordination and weakness of the lips, buccal muscles, and tongue, and is a strong indication of difficulty during the early oral phases.
- **Dehydration.** Dehydration is the most difficult finding to determine because its insidious onset causes a chronic condition. A generalized feeling of malaise, low-grade fever, decreased urine output, dry mouth, thick mucus, inability to clear pulmonary secretions, and decreased skin turgor are all symptoms of chronic dehydration.
• Problems at isolated, specific sites along the swallowing mechanism. The specific sites of dysphagia often can be determined early in the course of the disorder. As the disorder progresses, the problems usually become multiple and more generalized.