Dysarthria in Amyotrophic Lateral Sclerosis: A Systematic Review of Characteristics, Speech Treatment, and Augmentative and Alternative Communication Options

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This evidence-based review addresses intervention and management of dysarthria related to amyotrophic lateral sclerosis (ALS; Lou Gehrig's disease) and is part of a series of evidence-based systematic reviews sponsored by the Academy of Neurologic Communication Disorders and Sciences. A search of electronic databases (PsycINFO, Medline, and CINAHL) and hand search of relevant edited books yielded 713 articles on the topics related to characteristics, intervention, and management of the dysarthrias associated with ALS. This review summarizes the characteristics of dysarthria in ALS and appraises and summarizes findings from studies investigating speech treatment and augmentative and alternative communication (AAC) interventions for people with ALS. Findings include (1) well-documented characteristics of the progressive, mixed spastic and flaccid dysarthria; (2) consensus of expert opinion indicating the benefits of communication strategies, including speech supplementation and partner training for mild to moderate dysarthria; (3) a lack of evidence supporting the use of strengthening exercises for improving speech; (4) usefulness of monitoring of rates of speech to predict intelligibility declines and thus inform the timing of AAC intervention; (5) emerging evidence of the long-term usefulness of AAC systems; and (6) the influence of other factors, such as cognitive decline, that may affect the success of various interventions.

INTRODUCTION

In 1997, the Academy of Neurologic Communication Disorders and Sciences (ANCDS) established a committee to conduct a series of evidence-based systematic reviews related to dysarthria. These systematic reviews are intended for use in making clinical decisions about intervention for dysarthria in children and adults. Systematic reviews can be viewed as a process of translating evidence from both research literature and expert opinion into material that will help clinicians make management decisions. To date, six reviews have been completed and are available at the ANCDS's
website (http://www.ancds.org/). These reviews focus on various types of intervention: management of velopharyngeal, respiratory, or phonatory dysfunction; treatment of rate, loudness; or prosody; and speech supplementation. The current review is the first in a series of reviews focusing on neurologic conditions and the management of dysarthria in these specific populations.

THE CONDITION

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig’s disease, is a rapidly progressive degenerative disease involving the upper and lower motor neurons of both the brain and spinal cord (Mitchell & Borasio, 2007). It occurs more commonly in men than women with symptoms typically appearing between the ages of 40 and 70 years. Weakness is a usual early symptom with bulbar symptoms occurring initially in approximately 20 percent of cases (Wijesekera & Leight, 2009). The presence of dysarthria in individuals with ALS is both common and distressing. In a survey of people with ALS, the potential loss of speech is rated as the one of the worst aspects of the condition (Hecht et al., 2002).

Because the onset of symptoms is both insidious and variable and there are no biologic markers for the condition, early absolute diagnosis is difficult. To aid in diagnosis for both clinical and research purposes, a set of diagnostic criteria has been proposed and revised (Table 1). According to the El Escorial (Airlie Revision) diagnostic criteria (Brooks, Miller, Swash, & Munsat, 2000), the clinical level of certainty (made without pathological verification) can be categorized as clinically definite, probable, or possible.

The purpose of this systematic review is to evaluate the published literature related to dysarthria associated with ALS and interventions to treat or manage patients with this condition. The review is organized in order to address three clinically important questions: (1) What dysarthria characteristics are associated with ALS? (2) Is speech treatment beneficial? and (3) What augmentative and alternative communication (AAC) options are available when speech effectiveness is affected? A number of nonspeech factors that affect decisions about speech management are also discussed. Interested readers are directed to Palovcak et al. (2007) for a review of assessment and management of dysphagia associated with ALS.

METHODS

We searched the following electronic databases, chosen for their relevance to the topics of interest: PsycINFO, a database from the American Psychological Association covering 2450 journals (through May 2010); PubMed, a service of the National Library of Medicine with citations from medical and science literature (through December 2009); and CINAHL, covering 2900 journals (through December 2009). The initial searches were keywords paired with the terms “amyotrophic lateral sclerosis” (ALS) or “motor neuron disease” with “speech” or “dysarthria” or “communication.” In addition to these electronic searches, the authors conducted hand searches of relevant edited books in the area of dysarthria and ancestral searches of extant references (e.g., studies cited within an article or chapter). This search yielded 713 articles. To organize the review, the authors initially read each abstract to identify the topic(s), type of evidence (e.g., clinical trials, observational, case study, expert opinion) and closeness to target (i.e., “on target,” “closely related,” “somewhat related,” “not related”). Pertinent abstracts were

<table>
<thead>
<tr>
<th>TABLE 1. Diagnostic Criteria for Amyotrophic Lateral Sclerosis*</th>
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<tr>
<td>Clinically definite</td>
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<tr>
<td>Clinically probable</td>
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<tr>
<td>Clinically possible</td>
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</table>

*Region refers to the brainstem, cervical thoracic, or lumbosacral spinal cord levels. EMG = electromyographic; LMN = lower motor neuron; UMN = upper motor neuron. Adapted from Brooks, Miller, Swash, & Munsat, 2000.
selected, and complete texts were obtained for the articles related to the topics of speech characteristics, speech treatment, and AAC intervention. Inclusion criteria for citations and procedures for rating the strength of evidence are described here for each question.

RESULTS

What Dysarthria Characteristics Are Associated with Amyotrophic Lateral Sclerosis?

To answer this question, four topics are addressed: (1) the onset and progression of dysarthria, (2) physiologic impairment, (3) speech characteristics, and (4) the relationship of speech intelligibility with other variables. The following types of articles were included in the search: data-based descriptive studies, case studies, comparison of dysarthria in ALS to other types of dysarthria, and experimental manipulation of speech characteristics in ALS dysarthria.

Onset and Progression

The onset of dysarthria is insidious and may mimic other conditions such as adductor spasmodic dysphonia (Roth, Glaze, Goding, & David, 1996). In a study of consecutive cases (n = 111) seen in an outpatient speech clinic (Yorkston, Strand, Miller, Hillel, & Smith, 1993), the appearance of first speech symptoms varies widely from 33 months before diagnosis to 60 months after diagnosis. Although speech changes occur earlier in those with bulbar onset, functional changes in speech are noted in both bulbar- and spinal-onset groups. Furthermore, after symptoms appear, the rate of bulbar progression is similar for the bulbar- and the spinal-onset groups.

A range of early speech symptoms exists. Early laryngeal involvement is identified in a number of studies (Riddel, McCuailey, Mulligan, & Tandan, 1995; Watts & Vanryckeghem, 2001) and often documented by acoustic changes (Robert, Pouget, Giovanni, Azulay, & Triglia, 1999). At times, the acoustic changes preceded clinical symptoms (Tomik, Tomik, Partyka, Skladzien, & Szczudlik, 2007). Initial bulbar symptoms are variable and may include voice changes, velopharyngeal dysfunction, and tongue fasciculations. Extensive motor neuron loss is also reported before the onset of perceptual speech symptoms (Rosenfield, Viswanath, Herbrick, & Nudelman, 1991) because extensive motor neuron loss occurs before weakness appears.

Ten publications from 1979 through 2010 provide longitudinal information about progressive speech symptoms in ALS. See Table 2 for a description of these studies. Many of these (n = 7) are case studies or case series. Case reports document longitudinal changes in physiologic measures of vocal, velopharyngeal (Cha & Patten, 1989; Dworkin & Hartman, 1979), lingual (Leeper, Millard, Bandur, & Hudson, 1996), and lip and jaw function (Yunusova, Green, Lindstrom, Ball, Pattee, & Zinman, 2010). Changes in speech function are described acoustically (Ramig, Scherer, Klasner, Titze, & Hori, 1990; Seikel, Wilcox, & Davis, 1991). A moderate to strong correlation is suggested between motor (as measured by physiologic impairment) and speech function (Yorkston, Strand, & Hume, 1998). This relationship is not simple and is complicated by compensatory processes involving multiple adjustments made by the speaker (DePaul, Robbins, Abbs, & Brooks, 1999). Changes in movement, speaking, or diadochokineti rates precede decline in speech intelligibility (Ball, Beukelman, & Pattee, 2004b; Nishio & Niimi, 2000; Yorkston et al., 1993; Yunusova et al., 2010). Although both the rate of progression and pattern of impairment are variable, the levels of functional change are relatively predictable and have been described as a series of 10 levels of speech function that appear in Table 3 (Hillel & Miller, 1989; Yorkston, Miller, & Strand, 2004).

Physiologic Impairments

The underlying speech change associated with ALS is a mixed dysarthria. Early in the progression, either flaccidity or spasticity may predominate, but later, both occur (Duffy, 2005). Involvement of bulbar musculature is bilateral, but the pattern of involvement across speech subsystems varies across speakers. In addition, the cranial nerve involvement in ALS tends to be "bulbar" (i.e., involving nerves of the lower brainstem). Thus, upper face and ocular movements are typically spared until late in the course of the disease. Studies examining physiologic impairments associated with dysarthria in ALS universally show a pattern of decline as compared with a nondisabled
# Progression of Dysarthria

<table>
<thead>
<tr>
<th>Reference</th>
<th>Type of Study</th>
<th>Measures</th>
<th>Subject(s) with ALS</th>
<th>Time Frame</th>
<th>Brief Summary of Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dworkin &amp; Hartman (1979)</td>
<td>Case study</td>
<td>Neurologic examination</td>
<td>49-year-old man</td>
<td>6 months</td>
<td>Physiologic measures of tongue and velopharyngeal function decline over a 6-month period from 11 to 17 months after diagnosis.</td>
</tr>
<tr>
<td>Ramig et al. (1990)</td>
<td>Case study</td>
<td>Acoustic measures of voice</td>
<td>69-year-old man</td>
<td>6 months</td>
<td>Acoustic measures moved in to abnormal direction over time.</td>
</tr>
<tr>
<td>Seikel et al. (1991)</td>
<td>Descriptive</td>
<td>Temporal acoustic measures: VOT and vowel duration</td>
<td>Three patients</td>
<td>2 years</td>
<td>All types of motor neuron disorders demonstrated some degree of neutralization of the prevocalic VOT, target vowel duration, and postvocalic closure duration.</td>
</tr>
<tr>
<td>Yorkston et al. (1993)</td>
<td>Descriptive</td>
<td>Speech severity scale and speech intelligibility</td>
<td>110 (58 men, 52 women; 44 followed longitudinally)</td>
<td>Variable periods from 6 to 20 months</td>
<td>Functional changes in speech occurred in both bulbar- and spinal-onset speakers; the rate of progression varied considerably, but changes occurred earlier in the bulbar group.</td>
</tr>
<tr>
<td>Leeper et al. (1996)</td>
<td>Descriptive</td>
<td>Selected measures of vocal acoustics</td>
<td>Nine men and three women; five bulbar and seven nonbulbar onset</td>
<td>4 months</td>
<td>Positive correlation between rating of vocal roughness and jitter or shimmer and negative relationship between perceptual ratings and signal-to-noise ratio.</td>
</tr>
<tr>
<td>Yorkston et al. (1998)</td>
<td>Descriptive</td>
<td>Clinical examination</td>
<td>Five men and five women</td>
<td>Variable periods from 2 to 28 months</td>
<td>Trend toward worsening motor function; moderate to strong correlation between motor and speech function.</td>
</tr>
<tr>
<td>DePaul et al. (1999)</td>
<td>Case study</td>
<td>Long-term tracking speech movement, intelligibility, severity, swallowing</td>
<td>31-year-old man</td>
<td>9 years</td>
<td>Underlying physiology was differentially and disproportionately impaired, but natural compensatory processes involved multiple motor control adjustments toward functional outcomes.</td>
</tr>
<tr>
<td>Nishio &amp; Niimi (2000)</td>
<td>Case study</td>
<td>Rate, maximum repetition rates, and intelligibility</td>
<td>Case 1: 37-year-old man; case 2: 66-year-old woman</td>
<td>Case 1: 14 months; case 2: 7 months</td>
<td>Rate parameters (speaking and DDK) are more sensitive in reflecting early functional changes than intelligibility.</td>
</tr>
<tr>
<td>Watts &amp; Vanryckeghem (2001)</td>
<td>Case study</td>
<td>Laryngeal examination and vocal acoustics</td>
<td>72-year-old woman</td>
<td>6 months</td>
<td>Assessment of laryngeal dysfunction can assist diagnosis.</td>
</tr>
<tr>
<td>Yunusova et al. (2010)</td>
<td>Case series</td>
<td>Lip and jaw kinematics, speech intelligibility and rate</td>
<td>Three men, ages 44, 46, and 49 years</td>
<td>12, 5, and 7 months</td>
<td>Increased movement duration coincides with declines in speech intelligibility.</td>
</tr>
</tbody>
</table>

ALS = amyotrophic lateral sclerosis; DDK = diadochokinetic; VOT = voice onset time.
TABLE 3. Levels of Speech Function in Amyotrophic Lateral Sclerosis

Normal Speech Processes
10—Normal speech: Patient denies any difficulty speaking. Examination demonstrates no abnormality.
9—Nominal speech abnormality: Only the patient or spouse notices that speech has changed. Maintains normal rate and volume.

Detectable Speech Disturbances
8—Perceived speech changes: Speech changes are noted by others, especially during fatigue or stress. Rate of speech remains essentially normal.
7—Obvious speech abnormalities: Speech is consistently impaired. Rate, articulation, and resonance are affected. Remains easily understood.

Behavioral Modifications
6—Repeats messages on occasion: Rate is much slower. Repeats specific words in adverse listening situations. Does not limit complexity or length of message.
5—Frequent repeating required: Speech is slow and labored. Extensive repetition or a “translator” is commonly needed. Individual probably limits the complexity or length of messages.

Use of Augmentative Communication
4—Speech plus augmentative communication: Speech is used in response to questions. Intelligibility problems need to be resolved by writing or a spokesperson.
3—Limits speech to one-word response: Vocalizes one-word response beyond yes or no; otherwise writes or uses a spokesperson. Initiates communication nonvocally.

Loss of Useful Speech
2—Vocalizes for emotional expression: Uses vocal inflection to express emotion, affirmation, and negation.
1—Nonvocal: Vocalization is effortful, limited in duration, and rarely attempted. May vocalize for crying or pain.
X—Tracheostomy

Adapted from Yorkston, Miller, & Strand, 2004.

group. Most of these studies provide descriptive data about components of the speech production process, including:

• Tongue (Carrow, Rivera, Maudlin, & Shamblin, 1974; DePaul, Abbs, Caligiuri, Gracco, & Brooks, 1988; DePaul, Waclawik, Abbs, & Brooks, 1998; Dworkin, 1980)
• Larynx (Rosenfield et al., 1991; Strand, Buder, Yorkston, & Ramig, 1994; Tomik et al., 2007)
• Velopharynx (Delorey, Leeper, & Hudson, 1999)
• Lip and jaw (Yunusova et al., 2010)
• Interarticulator coordination (Weismer, Yunusova, & Westbury, 2003)

Speech Characteristics
The characteristics of speech in individuals with ALS were first described as part of the classic Mayo Clinic study (Darley, Aronson, & Brown, 1969, 1975). The speech of 30 patients with ALS was rated perceptually along 38 dimensions. Results suggest clusters of deviant dimensions associated with both spasticity and flaccidity, including articulatory and resonatory incompetence, phonatory stenosis or incompetence, and prosodic changes. The speech of those with ALS is characterized as slow and labored and marked by short phrases and intervals between words, defective articulation, a strained-strangled voice quality, hypernasality and mono pitch, and loudness (Duffy, 2005).

Since the classic work of Darley et al. (1975), a number of research groups have helped to document and refine the perceptual descriptions by examining the acoustic features of ALS dysarthria. Results suggest a variety of changes:

• Lengthening of segment duration (Tjaden & Turner, 2000)
• Voice onset timing disruptions (Caruso & Burton, 1987)
• Reduced spectral distinctiveness of fricative (Tjaden & Turner, 1997)
Increased vowel duration (Turner & Weismer, 1993)
- Reduced vowel space (Weismer, Jeng, Laures, Kent, & Kent, 2001)
- Flattened F2 slope (Kent, Kent, Weismer, & Martin, 1989; Mulligan et al., 1994; Weismer, Martin, Kent, & Kent, 1992)

Relationship of Speech Intelligibility to Other Variables

In addition to the perceptual and acoustic descriptions of ALS dysarthria, a growing literature seeks to understand the relationship between speech intelligibility and a variety of other factors. Thirteen such studies are summarized in Table 4. These studies suggest that changes in intelligibility are associated with physiologic factors that include changes in tongue force (Dworkin, Aronson, & Mulder, 1980) and slow movement rates (Langmore & Lehman, 1994; Yunusova et al., 2010). Changes in speech intelligibility are also associated with a variety of acoustic features, including a reduced F2 slope (Kent et al., 1989), abnormal trajectories in less intelligible speakers (Mulligan et al., 1994; Weismer et al., 1992), and smaller vowel spaces (Turner, Tjaden, & Weismer, 1995). Changes in speech intelligibility have been linked to changes in the production of speech sounds (phonetic contrasts) (Kent et al., 1990; Kent et al., 1992; Kent, Weismer, Kent, & Rosenbek, 1989). In a study of prosodic dysfunction, results indicated that speech intelligibility and naturalness may be separate factors, although both may be compromised in ALS dysarthria (Bunton, Kent, Kent, & Rosenbek, 2000). Finally, the relationship between a decline in speech intelligibility and self-reported ratings of communicative effectiveness has been examined (Ball et al., 2004b). Results indicated that although there is a positive relationship between intelligibility and effectiveness, even minor changes in intelligibility (declines in speech intelligibility to 90% or less) are associated with marked reduction in self-reported communicative effectiveness. Because intelligibility is often reduced in ALS dysarthria, the presence of compromised communicative effectiveness warrants referral for SLP services.

Clinical Characteristics of Dysarthria in Amyotrophic Lateral Sclerosis

- Type: Mixed flaccid-spastic dysarthria
- Progression: Progressive; ultimately leading to loss of intelligible speech.
- Variability: Variable physiologic impairments, particularly at onset. However, tongue and laryngeal impairments are common. The onset of dysarthria may precede other neurologic symptoms (as in bulbar onset) or be accompanied by limb symptoms (as in mixed or spinal onset).
- Pattern of change: Changes in motor function (e.g., movement and speaking rates) precede changes in speech intelligibility.

Research Recommendations

- More research is needed to understand the factors that prolong the usefulness of natural speech.
- Research is needed to understand the role of slowed speaking rate in ALS speech. For example, do speakers with ALS slow their speech as a compensatory mechanism in response to articulatory imprecision, or is the slower rate part of the physiologic changes characteristic of the dysarthria?
- Longitudinal studies are needed to refine our understanding of the predictors of changes in speech intelligibility.

IS SPEECH TREATMENT BENEFICIAL?

In this review, speech treatment is defined as a set of interventions designed to reduce impairment of the speech production mechanism or improve the quality of speech either through prosthetic or behavioral compensation. These are typically used when dysarthria is mild or moderate. With a small number of exceptions, described in the following sections, most of the literature reflects expert opinion. A number of general discussions of the management of ALS and the dysarthria associated with it are available (Francis, Bach, & DeLisa, 1999; Kuhnlein et al., 2008; Leigh et al., 2003; Tomik & Guiloff, 2008). Studies in the following sections were rated using guidelines published by the American Academy of Neurology (AAN) (Edlund, Gronseth, So, Franklin, & American Academy of Neurology, 2004). This classification scheme was
<table>
<thead>
<tr>
<th>Reference</th>
<th>Measures</th>
<th>Subjects</th>
<th>Brief Summary of Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dworkin et al. (1980)</td>
<td>Force transduction</td>
<td>19 individuals with ALS; 125 individuals without ALS</td>
<td>A high negative correlation between tongue force and severity of articulatory defect</td>
</tr>
<tr>
<td>Kent et al. (1989)</td>
<td>Word intelligibility and second formant slope</td>
<td>25 men with ALS; 10 women with ALS</td>
<td>Moderately high correlation indicates the F2 slope index is a useful acoustic measure of speech proficiency</td>
</tr>
<tr>
<td>Kent et al. (1989)</td>
<td>Single-word intelligibility</td>
<td>13 men with ALS</td>
<td>This test is designed to examine 19 acoustic–phonetic contrasts that are likely to be sensitive to dysarthric impairment and contribute significantly to speech intelligibility.</td>
</tr>
<tr>
<td>Kent et al. (1990)</td>
<td>Single-word intelligibility</td>
<td>25 men with ALS</td>
<td>Most disrupted phonetic features involved phonatory, velopharyngeal, place and manner of articulation for lingual consonants, and regulation of tongue height.</td>
</tr>
<tr>
<td>Weismer et al. (1992)</td>
<td>Acoustics</td>
<td>25 men with ALS; 15 control participants</td>
<td>In ALS speakers, those who were less than 70% intelligible produced distinctly more aberrant trajectory characteristics than subjects who were more than 70% intelligible and produced many trajectories that were essentially flat, or that had very shallow slopes.</td>
</tr>
<tr>
<td>Kent et al. (1992)</td>
<td>Phonetic features and acoustics</td>
<td>10 women with ALS</td>
<td>Phonetic changes pertained to velopharyngeal valving, lingual function for consonant contrasts of place and manner, and syllable shape.</td>
</tr>
<tr>
<td>Mulligan et al. (1994)</td>
<td>F2 transition rate</td>
<td>Seven dysarthric with ALS; seven nondysarthric with ALS</td>
<td>A relationship between the F2 transition rate and single-word intelligibility was noted for patients with moderate to high intelligibility, but at lower levels of intelligibility, the F2 rate reached a plateau despite continued decline in intelligibility.</td>
</tr>
<tr>
<td>Langmore &amp; Lehman (1994)</td>
<td>Physiologic variables</td>
<td>14 individuals with ALS; 15 control participants</td>
<td>The tongue was generally the most affected structure in all ALS groups. Perceived severity of dysarthria was more highly correlated with the measures of repeated contraction rate than with the measures of strength, suggesting that more severe dysarthria may be largely attributable to slower movement of the orofacial structures until substantial muscle strength has been lost.</td>
</tr>
<tr>
<td>Turner et al. (1995)</td>
<td>Acoustic</td>
<td>Nine individuals with ALS; nine control participants</td>
<td>Dysarthric speakers exhibited smaller vowel space areas and less systematic changes in vowel space as a function of speaking rate. Vowel space was found to account for 45% of the variance in speech intelligibility.</td>
</tr>
<tr>
<td>Study</td>
<td>Methodology</td>
<td>Sample Description</td>
<td>Findings</td>
</tr>
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<tr>
<td>Bunton et al. (2000)</td>
<td>Perceptual ratings of prosody and acoustics</td>
<td>Five control participants; 10 with ALS (high and moderate), including five with cerebellar disease</td>
<td>Dysprosody was assessed with perceptual ratings and acoustic measures pertaining to the regulation of duration, F0, and intensity within tone units of conversational samples. Intelligibility reduction and prosodic disturbance were not necessarily equally impaired in all subjects, and it was concluded that these are complementary indices of severity of dysarthria.</td>
</tr>
<tr>
<td>Weismer et al. (2001)</td>
<td>Scaled speech intelligibility and selected acoustic variables</td>
<td>ALS (five men and five women); PD; control participants</td>
<td>Temporal variables typically differentiated the ALS group, but not the PD group, from the control participants, and vowel spaces were smaller for both neurogenic groups compared with control participants but only significantly so for the ALS speakers.</td>
</tr>
<tr>
<td>Ball et al. (2004)</td>
<td>Speech intelligibility, rate, and self-reported communicative effectiveness</td>
<td>10 women and 15 men with ALS</td>
<td>Although there is a positive relationship between intelligibility and self-reported effectiveness, decreases in communication effectiveness are noted with minor changes in speech intelligibility.</td>
</tr>
<tr>
<td>Yunusova et al. (2005)</td>
<td>Acoustics</td>
<td>Seven individuals with ALS; three with PD; 10 control participants</td>
<td>Linguistic or acoustic variables that predict across-speaker variations in speech intelligibility may not function in the same way when within-speaker variations in intelligibility are considered.</td>
</tr>
<tr>
<td>Yunusova et al. (2010)</td>
<td>Kinematics</td>
<td>Three men with ALS, ages 44, 46, and 49 years</td>
<td>Increased movement duration coincide with declines in speech intelligibility.</td>
</tr>
</tbody>
</table>

ALS = amyotrophic lateral sclerosis; PD = Parkinson’s disease.
TABLE 5. American Academy of Neurology Classification of Evidence for Therapeutic Intervention

Class I: A randomized, controlled clinical trial of the intervention of interest with masked or objective outcome assessment in a representative population. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences.

Class II: A randomized controlled clinical trial of the intervention of interest in a representative population with masked or objective outcome assessment that lacks some criteria for class I or a prospective matched cohort study with masked or objective outcome assessment in a representative population that otherwise meets class I criteria. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences.

Class III: All other controlled trials (including well-defined natural history control participants or patients serving as own control participants) in a representative population, where outcome is independently assessed, or independently derived by objective outcome measurement.

Class IV: Studies not meeting class I, II, or III criteria, including consensus or expert opinion.

Adapted from Edlund, So, & Franklin, 2004.

developed to rate level of evidence in therapeutic interventions (Table 5).

There is little research examining speech intervention effectiveness in ALS. Not only is there a complete absence of randomized controlled trials (AAN class I and II evidence), but we found only three experimental studies (all class III evidence) that examine specific therapeutic interventions for dysarthria in ALS. These are discussed below. In addition, one article reports on the global benefits of multidisciplinary intervention. Multidisciplinary intervention (including speech treatment) was shown to improve quality of life in individuals with ALS (Van den Berg et al., 2005).

Compensating for the Impairment

Prosthetic compensation for velopharyngeal dysfunction was reported in two studies (one class III and one case study) (Esposito, Mitsumoto, & Shanks, 2000; Roth, Roburka, & Workinger, 2000). Roth et al. (2000) report that intelligibility improved from 16% to 71% in a patient with ALS after placement of a palatal lift. The authors found that the palatal lift was beneficial throughout 18 months of disease progression. In a retrospective study, Esposito et al. (2000) report that 21 patients (84%) treated with a palatal lift demonstrated improvement in their dysarthria, specifically in reduction of hypernasality, with 19 (76%) benefiting at least moderately for 6 months. Of the 10 patients treated with a combination palatal lift and an augmentation prosthesis that lowers the palate, six (60%) demonstrated improvement in articulation. A majority of patients indicated they spoke with less effort when wearing the prosthesis.

Although there is a consensus of expert opinion that behavioral strategies to compensate for mild to moderate dysarthria are effective, only one research study provides data-based evidence for this opinion. Murphy (2004a) reported qualitative data related to types of communicative strategies that people with ALS and their communicative partners reported to be useful. These strategies include low-technology AAC strategies such as alphabet charts, conversational strategies involving the communication partner, speaking strategies such as repetition, and nonverbal strategies such as gestures.

Exercise

Literature examining the efficacy of exercises to strengthen oropharyngeal musculature in ALS is limited to two case studies (Dworkin & Hartman, 1979; Watts & Vanryckeghem, 2001). In both cases, the dysarthria worsened after the intervention. Watts and Vanryckeghem (2001) reported on the effects of speech and voice intervention for a woman with bulbar ALS. Initially, the Lee Silverman Voice Treatment (LSVT) was applied with appropriate speech breathing patterns and voice focus with resonant therapy. The LSVT was ineffective. For this speaker, it resulted in reduced voice quality and the appearance of ventricular phonation. After use of the LSVT, voice-focused treatment and glottal fry techniques resulted in reduced ventricular phonation, but the speaker's voice subsequently returned to a very low volume. Voice therapy was phased out. Articulation treatment included oral motor motility and strengthening exercises. Intelligibility continued to decline despite the strengthening exercises. Intensive
voice and articulation therapy was ineffective. Similarly, Dworkin & Hartman (1979) reported the case of a subject with bulbar ALS who showed no improvements after intensive intervention that included tongue-strengthening exercises.

Clinical Findings

- Strong consensus of expert opinion that behavioral strategies (including speech supplementation, partner support, and other speaking and nonverbal strategies) are useful for individuals with mild to moderate dysarthria.
- Limited evidence that prosthetic management of velopharyngeal impairment may be beneficial for some for a limited period of time.
- No evidence is available to support the use of strengthening exercises.

Research Recommendations

- There is a need for higher levels of research evidence (e.g., AAN levels I, II, or III) investigating the general effectiveness of behavioral strategies to compensate for dysarthria in ALS, including the type and timing of such strategies.
- There is a need for higher levels of research evidence examining factors that may prolong the usefulness of natural speech.

WHAT COMMUNICATION OPTIONS ARE AVAILABLE WHEN SPEECH EFFECTIVENESS IS AFFECTED?

Augmentative and alternative communication is a term referring to any type of communication, other than natural speech or writing, that meets a person’s needs on a temporary or permanent basis (Beukelman & Mirenda, 2005). AAC intervention encompasses communication strategies that require little to no external aids, such as gestures and sign language, as well as portable electronic (i.e., high-tech) speech-generating devices (SGDs) with multiple modes of access and synthesized voice output for those with speech impairment and limited limb control. Use of AAC interventions, including SGDs, is recognized as the standard of care for the treatment of individuals with speech-related functional losses caused by ALS.

A number of factors have spurred increasing interest in the applications of AAC systems. First, the majority of people diagnosed with ALS will lose their ability to speak (Ball, Beukelman, Ullman, Maassen, & Pattee, 2005) and to write (Birbaumer et al., 1999; Dobkin, 2007), making them obvious candidates for AAC intervention. Second, advances in technology, which are described later, have allowed access to AAC systems for those with even profound loss of movement. Third, other medical interventions, such as percutaneous endoscopic gastrostomy (PEG) and noninvasive ventilation, have prolonged the survival of people with ALS.

Finally, funding of AAC devices through Medicare has increased access to this technology (Medicare Implementation Team, 2001).

Use of the search term “augmentative and alternative communication” was added to the original search. This search resulted in 44 articles related to both AAC and ALS in the following categories (number of articles):

- General description of palliative care (4)
- General description of communication management (8)
- Observational studies of clinical populations (8)
- Survey of speech-language pathologists (SLPs) or caregivers (4)
- Experiences of people with ALS (interviews or surveys) (6)
- Case reports (6)
- Description of specific devices (8)

Rating of levels of evidence in the area of assistive technology poses a number of unique challenges that are not experienced in other areas such as appraisal of pharmaceutical studies (Johnston, Schere, & Whyte, 2006). For example, blinding is not possible because use of devices is visible. Furthermore, it is no more reasonable to test whether an AAC system is effective for someone who cannot speak than it is to test the efficacy of parachutes to reduce mortality in people jumping from airplanes (Smith & Pell, 2003). Rather, it is more reasonable to ask questions about patterns of use and the satisfaction of the user. Based on the review of this literature, a number of topics are reviewed below, including the pattern of use, the timing of critical management decisions, and future directions. Most are class IV evidence (Edlund et al., 2004), representing expert opinion, case studies, uncontrolled studies, or those not meeting criteria for class III or higher. Some represent class III evidence, which includes controlled research designs without randomization or blinding.
Patterns of Augmentative and Alternative Communication System Use

The patterns of use are described in a number of studies. Many of these studies address the types of communication function reported by users of AAC systems. People with ALS need to communicate about the same topics and for the same reasons as anyone else. They communicate to stay connected and interact socially (Brownlee & Palovcak, 2007; Dobkin, 2007; Fried-Oken et al., 2006; Mitsumoto & Rabkin, 2007; Yorkston et al., 1993); to talk about their families (Todman & Lewins, 1996); to maintain employment (Ball, Beukelman, & Pattee, 2004a; McNaughton, Light, & Arnold, 2002; McNaughton, Light, & Groszyk, 2001; Zeitlin, Abrams, & Shah, 1995); to interact with health care providers and thus maintain control of their care (Brownlee & Palovcak, 2007; Yorkston et al., 1993); to reduce anxiety (Brownlee & Palovcak, 2007); and to take care of basic needs, especially when they are no longer able to take care of some things themselves (Dobkin, 2007; Fried-Oken et al., 2006; Harris & Goren, 2009). In addition to specific communicative function, AAC systems allow people with ALS to retain their autonomy and decrease their dependence on others for communication (Nijboer et al., 2008). Valuing communication and striving to help people with ALS to communicate effectively fosters hope and allows the person to express fear, pain, and grief related to the disease (Centers, 2007). Communication through AAC systems becomes more important in all of these areas as the person’s disability increases (Zeitlin, 1995), and use of AAC strategies can decrease the anger and frustration that grows when an individual is losing the ability to communicate through natural speech (Murphy, 2004a).

The duration of AAC system use has also been documented in a small number of studies. In an early report of 33 people with ALS (1988–1996), the mean duration of use was 14 months (Mathy, Yorkston, & Gutmann, 2000). In a somewhat later study of 45 participants, SGD use was 25 months for those with bulbar onset and 34 months for those with spinal onset of ALS. The length and initiation of use depended on several factors, including the timing of initial referral for AAC services and use of invasive ventilation. Medical interventions that prolong life (e.g., feeding tube placement or noninvasive ventilation) may influence the duration of AAC use, making the need for more high-tech systems even greater in the middle and end stages of the disease (Ball, Beukelman, Anderson, Bilyeu, Robertson, & Pattee, 2007; Beukelman, Fager, Ball, & Dietz, 2007).

Patterns of use change over time as the condition progresses. Alphabet supplementation, a strategy in which the speaker points to the first letter of each word while speaking it to increase speech intelligibility (Hanson, Yorkston, & Beukelman, 2004), may allow speakers to communicate using natural speech longer than they would have been able to without it. Use of alphabet supplementation accomplishes two important things. First, it extends the time that natural speech can be used for communication. It may also ease the transition to alternative communication systems because pointing to first letters is a first step to spelling entire words to communicate, as in the use of a low-tech alphabet board or a high-tech SGD.

People with ALS may initially rely on low-tech options such as writing messages or using a laminated alphabet board to spell out messages or high-tech options such as spelling messages on an SGD. As voluntary muscle control of hands and arms is gradually lost and speech deteriorates, the person may need to transition to alternative access methods such as eye gaze or infrared mouse emulation. Doyle and Phillips (2001) described the contour of AAC use changing from low-tech and unaided communication in the early stages, transitioning to high-tech systems during the middle stage, and sometimes back to low-tech systems in the end stage. Holmøy and Worren (2006) found that patients in Norway used a variety of AAC systems from low to high tech, often combining them for the best effect.

Critical Management Decisions in Augmentative and Alternative Communication Intervention

Decisions about the timing of intervention and acceptance of devices are critical for successful management of people with severe or profound dysarthria in ALS. There is strong consensus of expert opinion that early AAC intervention is preferred over delaying intervention until speech is no longer intelligible. The latter tends to lead to implementing AAC in a climate of crisis (Gelinas, 1997; Murphy, 2004b; Nordness, Ball, Fager, Beukelman, & Pattee, 2010; Yorkston et al., 2004). Nordness et al. (2010) suggest a proactive decision-making approach to acquiring AAC technology. They reported the most common reason for
a delay in AAC assessment was lack of a physician's referral. Knowing when to discuss sensitive issues and treatment decisions, such as loss of natural speech and the idea of AAC, remains challenging (Mandler, Anderson, Miller, Clawson, Cudkowicz, & Del Bene, 2001). People with ALS and their family members are often not interested in facing the loss of their speech while it is still functional and thus may not wish to participate in the evaluation process or even learn about AAC options (Ball et al., 2005; Nordness et al., 2010).

Clinical opinion has long suggested that decreases in speaking rate precede changes in speech intelligibility in ALS and thus may inform the timing of AAC assessment and intervention (Yorkston et al., 1993). More recent observational studies of large clinical populations have also identified slowed speaking rate, altered voice quality, and low communication effectiveness ratings as the strongest early predictors of impending intelligibility declines in speakers with ALS (Ball, Willis, Beukelman, & Pattee, 2001). Recent physiologic studies have also shown that changes in movement rates precede changes in speech intelligibility (Yunusova et al., 2010).

Ball, Beukelman, and Pattee (2002) suggest referral guidelines based on habitual speaking rate and recommend that the AAC assessment be initiated when speaking rate decreases to 125 words per minute (wpm) compared with a typical speaking rate of 180 to 200 wpm on the Sentence Intelligibility Test (Yorkston, Beukelman, Hakel, & Dorsey, 2007). If progression proceeds in a typical fashion, then speech intelligibility will soon decline. The investigators went on to develop a protocol to regularly measure speaking rate via the telephone that proves to be a useful assessment strategy for people who live far from regional clinics (Ball et al., 2005). Ongoing monitoring of speaking rate is a way to identify changes and to introduce the possibility of using AAC technology without overwhelming people with ALS and their families with premature decisions about AAC technology (Ball et al., 2004a; Nordness et al., 2010). Taken together, these findings represent a clinically important advance in monitoring and predicting speech changes that should inform the timing of the AAC evaluation and timely acquisition of the right system. Details about typical AAC assessment procedures can be found elsewhere (Ball, Beukelman, & Bardach, 2007).

Acceptance of AAC systems has been studied in many clinical populations. Ball et al. (2004a) interviewed 50 people with ALS in their regional clinic and found that the vast majority (90%) accepted the AAC option immediately. People who first declined but later accepted AAC (delayed acceptance rate, 6%) identified negative reactions of family members or physicians as the reason they initially refused AAC. Reports of non-acceptance of AAC systems are generally based on small samples (Murphy, 2004a; Perry, Gowel, & Rose, 1981; Zeitlin et al., 1995) or single case descriptions (DePaul & Kent, 2000; DePaul et al., 1999). Studies examining the reasons for non-acceptance identify cognitive decline (Ball et al., 2004a; Zeitlin et al., 1995), finances (Zeitlin et al., 1995), the desire for more intimate partner involvement in the interaction (Murphy, 2004b), and partner attitudes (Ball et al., 2004a; DePaul & Kent, 2000; Doyle & Phillips, 2001). Because attitudes of family members and health care providers greatly influence willingness to accept an AAC system, a study was undertaken to examine listener attitudes. Listeners preferred AAC systems to dysarthric speech that is difficult to understand (Richter, Ball, Beukelman, Lasker, & Ullman, 2003).

**Future Directions in Augmentative and Alternative Communication Intervention**

A number of directions for future development have been suggested by people with ALS who use AAC systems (McNaughton et al., 2001). These include decreasing the learning demands of the system; easy online instructions; better synthesized speech; and lighter, smaller, more attractive devices. Increasing conversational rate remains an important goal for users (Todman & Lewins, 1996).

Improved access to computers and AAC systems is also an important research and development focus. Eye tracking access is available in a growing number of communication systems and works with a special camera that tracks the movement of the pupil and glint of the eye, translating it into cursor control (Beukelman et al., 2007; Harris & Goren, 2009). Because eye control is typically retained longer than other muscle control in individuals with ALS, this is an especially useful mode of access to AAC technology and should be refined in order to work better for a greater number of people.

Brain–computer interface (BCI) technology circumvents the need for any muscle control by measuring and translating brainwaves.
into computer control that can, in turn be used to select letters on an onscreen keyboard or environmental controls (Birbaumer et al., 1999; Daly & Wolpaw, 2008; Dobkin, 2007; Hinterberger, Veit, Wilhelm, Weiskopf, Vatine, & Bierbäumer, 2005; Iversen, Koyama, Ohno, Ishihara, Nagumo, & Kawamura, 2008; Nijboer et al., 2008; Sellers & Donchin, 2006). Daly & Wolpaw (2008) reported word processing rates of 2 to 4 wpm with this access method, and although slow, they emphasized that restoring the ability of independent communication can have an important impact on quality of life for the people with ALS and their caregivers.

Voice message banking is a way for people facing permanent loss of speech to record and preserve messages in their own voices through digital recording (Costello, 2009). The possibility of preserving one’s speech before losing it is of great potential use, especially if speakers are evaluated for an SGD early enough in the course of their disease to undertake the recording before their speech becomes unintelligible. Voice banking is accomplished through different procedures described by Costello (2009) in his work in palliative care for children.

Clinical Findings

• Early speech-language assessment to establish baseline of speaking rate is important.
• Ongoing monitoring of speaking rate is essential to inform timing of AAC assessment and intervention.
• Ongoing monitoring of communication needs allows the clinician to respond to communication changes in a timely way.
• Voice banking may be undertaken while speech is still intelligible.
• Alternative forms of access to SGDs can help people with ALS continue to communicate even when motor control is extremely limited.

Research Recommendations

• Expand investigations of ease and effectiveness of system access for those with profound motor problems, including BCI and eye gaze technology.
• Expand investigations of consumer satisfaction and usage patterns.
• Investigate changes in quality of life status that may be related to AAC intervention.
• Investigate the role of AAC use in end-of-life communication and decisions.
• Apply well-controlled research designs to systematically compare measurable variables such as the type and extent of training or various system features.
• Investigate the influence of natural language processing on text and message preparation in AAC devices to increase the communication rate.
• Explore application of mobile, multimedia consumer technologies to support the communication needs of people with ALS.
• Compare and contrast voice-banking procedures and investigate outcomes such as cost-benefit analyses of the trade-offs between the time necessary to do the recording and attendant fatigue caused by the recording process and the benefits of preserving specific messages recorded in one’s own voice.

OTHER FACTORS TO CONSIDER

A variety of additional factors affect the management of dysarthria in individuals with ALS. These include cognition, pseudobulbar affect, apraxia of speech (AOS), respiration, and saliva control, which are discussed below.

Cognition

Many people diagnosed with ALS present with normal cognitive skills. However, a subset of this population experiences cognitive decline. As many as 15% of people with ALS develop a concomitant frontotemporal dementia (FTD) (Ringholz, Appel, Bradshaw, Cooke, Mosnik, & Schulz, 2005). In a larger percentage of cases, however, there are more subtle impairments of executive functions (e.g., deficits in memory, attention, judgment) that do not meet criteria for FTD. Evidence of executive function deficits has been reported in 50% to 75% of the patients examined, most with bulbar ALS (Abe, Juirimura, Yoyooka, Sakoda, Yorifuji, & Yanagihara, 1997; Lomen-Hoerth, Murphy, Langmore, Kramer, Olney, & Miller, 2003; Sterling et al., 2010). In a recent meta-analysis of cognition in ALS, 16 studies including 554 patients with ALS were analyzed.
(Raaphorst, de Visser, Linssen, de Haan, & Schmand, 2010). Although movement rates (that may have impacted response latency) may have influenced performance despite attempts to correct for it, conclusions suggested that cognitive problems in ALS may be greater than was previously thought. People with bulbar-onset ALS have more consistently demonstrated deficits on neuropsychological tests examining executive function skills, attention, and memory than those with spinal-onset ALS (Schreiber et al., 2005; Sterling et al., 2010). Cognitive impairment has not been shown to deteriorate over time in conjunction with decline in motor ability. It is possible that other factors might contribute to mild deficits with memory and attention, including medications, chronic hypoventilation, pain, fatigue, or depression. Although research has demonstrated that cognitive decline is more prevalent than previously believed, mild cognitive impairment does not typically interfere substantially with the ability to carry out speech treatment strategies (Ball et al., 2007). However, for the subset of persons with ALS who develop more significant dementia, the treatment approach must be modified.

Apraxia of Speech

In addition to the concomitant presentation of the dysarthria associated with ALS and cognitive deficits, evidence is beginning to suggest that AOS is another co-occurring condition. Duffy, Peach, and Strand (2007) evaluated a subset of seven patients identified with motor neuron disease and AOS from a larger set (n = 80) of patients with varied degenerative diseases concomitant with AOS. All patients in the smaller group presented with speech characteristics consistent with AOS (e.g., distorted substitutions or difficulty with sequential motion rates) as well as dysarthria, with the AOS characteristics being the more prominent features in four of the cases. The authors suggest that, in some cases, the presence of AOS characteristics may be an early sign of ALS.

Pseudobulbar Affect

Emotional lability (pseudobulbar affect) is a frequent symptom that affects one’s ability to control laughter or crying. Within the context of ALS, it has been correlated with bulbar involvement (Newson-Davis, Abrahams, Goldstein, & Leigh, 1999). Pseudobulbar affect is believed to be a result of involvement of bilateral corticobulbar tracts in the subcortical white matter and brainstem that result in a loss of inhibitory control over emotional responses (Montgomery & Erickson, 1987). When severe, lability is disruptive, especially when communicating with those who are not aware of the nature of the problem.

Respiration and the Need for Assistive Ventilator Devices

Respiratory impairment eventually develops for most people diagnosed with ALS. This is of concern to SLPs because it may interfere with speech production or necessitate long-term AAC use if the patient chooses noninvasive or invasive ventilation. It is essential that intervention pertaining to communication be considered within the context of respiratory impairments and intervention. The AAN’s recent practice parameter update for ALS (Miller et al., 2009) recommends routine monitoring of respiratory function. However, it should be noted that no respiratory measure has shown predictive power in those with primarily bulbar weakness. AAN class I and class III evidence supports use of noninvasive ventilation (NIV) (e.g., bilevel positive airway pressure) to treat respiratory insufficiency in people with ALS. NIV has been shown to slow the rate of respiratory decline and lengthen survival. It may also contribute to improving quality of life in the face of this degenerative disease. However, in those with primarily bulbar dysfunction, NIV has not been as effective as it is for those with primarily limb involvement (Farrero, Prats, Povedano, Martinez-Matos, Manresa, & Escarrabill, 2005). Compliance with NIV use has been reportedly lower in those with concomitant frontotemporal dysfunction and in those with bulbar dysfunction, but NIV use has been correlated with use of PEG and speech devices (Miller et al., 2009).

Saliva Control

Saliva management and secretion clearance difficulties are common in ALS as bulbar involvement progresses beyond the mild stages. Sialorrhea occurs as a result of impaired ability of the oral muscles to seal the lips, transport saliva to the back of the mouth, and swallow; it is not a result of overproduction of saliva (Andersen et al., 2005).
When severe, it may interfere with the production of speech. For those who are unable to benefit from saliva management strategies, including medication management, AAN practice parameter guidelines suggest that Botox injections or low-dose radiation to the salivary glands (or both) may effectively treat sialorrhea (Miller et al., 2009).

**SUMMARY**

This systematic review summarizes the characteristics of dysarthria associated with ALS and evidence related to speech and AAC intervention as well as additional factors that may influence management decisions. The speech characteristics of ALS dysarthria are well documented to be a mixed flaccid–spastic dysarthria with a progressive course leading eventually to loss of speech. Tongue and laryngeal impairments are common early symptoms. As the condition progresses, all speech components become involved. Change in speech and movement rates precede changes in speech intelligibility.

Much less research emphasis has been placed on documenting the effectiveness of speech intervention in people with ALS. There is strong consensus of expert opinion that compensatory strategies such as alphabet supplementation to maintain communication are useful in mild to moderate dysarthria. Limited evidence exists suggesting that prosthetic management of the velopharyngeal system may be helpful to some for a limited period. No evidence is available to support strengthening exercises to improve speech in individuals with ALS.

A variety of AAC devices are available when speech effectiveness declines. Consensus of expert opinion and observational data support the following practices: early assessment and monitoring of speaking rate to inform the timing of AAC intervention, thorough training of both the person with ALS and his or her communication partners in use of the device and communication strategies, and continued follow-up to monitor changing communication needs and abilities. All communication intervention should be carried out within the context of cognitive and respiratory status. Because most of the evidence reported in this review is based on expert opinion and class IV evidence, higher level studies using more rigorous research design are needed to address many issues related to the management of dysarthria in individuals with ALS.

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**REFERENCES**


DYSARTHRIA IN AMYOTROPIC LATERAL SCLEROSIS

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Masses that Arise within or Encroach upon the Parapharyngeal Space

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Abnormally appearing parapharyngeal space (PPS) tissues may result from many different benign and malignant pathological processes. When confronted with suspicious conditions involving this complex region of the head and neck, clinicians may wish to adhere to a detailed, systematic, and comprehensive set of examination and treatment procedures to ensure appropriate diagnoses and successful treatment outcomes. This paper presents the differential workup of four patients who presented with suspicious masses that either directly or indirectly encroached upon the PPS. A suggested algorithm is introduced to facilitate accurate identification and treatment of abnormalities that involve the PPS and its anatomically proximal tissue network.

INTRODUCTION

The parapharyngeal space (PPS) resembles an inverted pyramid, with the narrow apical portion in the general vicinity of the greater horns of the hyoid bone and the broad basilar segment formed by components of the skull base. This potential space is one of several connective tissue planes of the head and neck, largely composed of deep layers of cervical fascia. It can be geometrically divided into three anatomical regions: (1) a wide masticator or prestyloid space enveloping several structures, including the inferior margin of the mandible, pterygoid, temporalis and masseter muscles, and deep portion of the parotid gland; (2) a thin region interspersed between the buccopharyngeal fascia and stylopharyngeal aponeurosis; and (3) the retrostyloid (i.e., carotid sheath) space located posterior to this aponeurosis (Allison, Van der Wall, & Snow, 1989; Bass, 1982; Shoss, Donovan, & Alford, 1985; Som, Biller, & Lawson, 1981; Som, Biller, Lawson, Sacher, & Lanzieri, 1984; Work, 1989). The internal carotid arteries (ICAs); internal jugular veins; and cranial nerves IX, X, XI, and XII all course through this third PPS compartment.

Whereas infectious and inflammatory conditions represent the vast majority of all pathological processes involving the PPS, benign (75%) and malignant (25%) neoplasms may affect any structure inherently contained within this space. These incidence data are especially true in the adult population. In the pediatric setting, virtually all presenting PPS abnormalities are caused by an underlying abscess. In both adults and children, very early PPS pathologies may be serendipitously discovered on routine physical examination without clinically significant associated symptoms. With progression of the underlying condition, signs and symptoms of odynophagia, oropharyngeal bulging,
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Editorial

Pathography of Love
Leonard L. LaPointe, Ph.D.

Articles

Botulinum Neurotoxin for Failed Tracheoesophageal Speech: Phonatory and Functional Voice Outcomes
Nadine R. Lawson, Bach. App. Sci (Speech Pathology),
Elizabeth C. Ward, BSpThy (Hons), Grad Cert Ed, Ph.D.,
Natalie J. Duncan, Bach Speech Pathology (Hons),
Malcom Baxter, M.B.B.S., FRACS, Andrew Sizeland, Ph.D., FRACS,
and Andrew J. Hughes, M.B.B.S., M.D., FRACS

Dysarthria in Amyotrophic Lateral Sclerosis:
A Systematic Review of Characteristics,
Speech Treatment, and Augmentative
and Alternative Communication Options
Elizabeth K. Hanson, Ph.D., Kathryn
M. Yorkston, Ph.D., and Deanna Britton, Ph.C.

Masses that Arise within or Encroach upon the
Parapharyngeal Space
James P. Dworkin, Ph.D., Richard
T. Klapchar, D.O., and David N. Madgy, D.O.

Multimodal Communication Training in Aphasia:
A Pilot Study
Mary Purdy, Ph.D., and Julie A. Van Dyke, Ph.D.