Assessment and Management of Communication and Swallowing in Patients with ALS
Insights from SLPs at Hospital For Special Care’s Neuromuscular Clinic.
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ALS = Amyotrophic Lateral Sclerosis

Amyotrophic - means “without nourishment to muscles” and refers to the loss of signals nerve cells normally send to muscle cells.
Lateral - means “to the side” and refers to the location of the damage in the spinal cord.
Sclerosis - means “hardened” and refers to the hardened nature of the spinal cord in advanced ALS.

ALS in Connecticut
• 200 – 250 people in CT currently have ALS.
• At HFSC we follow ~ 160 patients with ALS.
• 67 new ALS patients at HFSC ALS Clinic in 2009.
• 78 new ALS patients at HFSC in 2010 currently (likely due to increased referrals, not due to increased number of patients with ALS).

ALS Epidemiology
• Worldwide Incidence 0.86 to 2.4 per 100,000 per year (McGuire, V. & Nelson, L. M., 2006)
• US Incidence 2 per 100,000 per year
• Median survival rate is 3 years resulting in a prevalence rate of 6 per 100,000 per year.
• Onset typically occurs between 30-60 years of age
• Male to Female Ratio of 1.6:1

What is ALS? A Review.
• Lou Gehrig’s Disease
• Progressive
• Degenerative
• Upper and Lower Motor Neuron Disease
• Heterogeneous presentations
• No known cause
• No known cure

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Motor Neurons

ALS affects motor neurons that send impulses from the brain to the muscles of the body.

Therefore, the senses of touch, taste, sight, smell and hearing are not affected in pure motor neuron disease.

Review of Neuroanatomy

- UMN – neurons that have cell bodies in the brain and synapse on lower motor neurons.
- LMN – neurons that have cell bodies in the cranial nerve nuclei or the anterior horn of the spinal cord and synapse on muscle.

Motor Neuron Damage

<table>
<thead>
<tr>
<th>Upper Motor Neurons</th>
<th>Lower Motor Neurons</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spasticity or Increased Tone</td>
<td>Weakness of Skeletal &amp; Bulbar Muscles (moderate – severe)</td>
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<td>Weakness (mild-moderate)</td>
<td>Hypotonia</td>
</tr>
<tr>
<td>Clumsiness or Loss of Dexterity</td>
<td>Hyporeflexia</td>
</tr>
<tr>
<td>Hyperreflexia</td>
<td>Fasciculations</td>
</tr>
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<td>Pathological Reflexes</td>
<td>Muscle Atrophy or Wasting</td>
</tr>
<tr>
<td>Pseudobulbar Affect (Emotional Lability)</td>
<td>Cramps</td>
</tr>
</tbody>
</table>

Bulbar vs. Spinal Musculature

- **Bulbar Muscles**
  - Innervated by cranial nerves exiting from the brainstem
  - Control speech and swallowing musculature
- **Spinal Muscles**
  - Innervated by spinal nerves exiting the spinal cord
  - Control limb, trunk and respiratory musculature

Cranial Nerve Examination

- When you understand the etiology, individualized treatment plans are easier to develop and make sense.
- More than an oral mechanism exam that looks at the structure and function of the articulators, larynx and respiratory system. A Cranial Nerve Exam answers the questions WHY and WHERE?

Cranial Nerve Exams and ALS

25% of patients with ALS develop bulbar symptoms initially (dysarthria & dysphagia)

In LMN-predominant bulbar ALS, lingual weakness, atrophy and fasciculations typically are the first to present.

Many bulbar-ALS patients are incorrectly diagnosed initially or not referred early to Neurologist by their PCPs.
Resources For Further Study

• http://anatomy.med.umich.edu/nervous_system/antneck_tables.html

You can also find plenty of decent videos illustrating cranial nerve examinations on You Tube!

Normal Tongue Bulk Vs. Atrophied Tongue due to ALS

Tongue Atrophy - ALS Normal Tongue Bulk

Tongue Atrophy in a 46-year old man with ALS

Primary Lateral Sclerosis (PLS)

• Slowly progressive degeneration of the UMs in the brain and spinal cord that control voluntary movements that may develop into ALS should LMN signs and symptoms appear.
• Causes gradually disabling spasticity and weakness.
• Does not affect LMNs.
• Muscles are not directly affected (no muscle wasting/atrophy or fasciculations).

Spectrum of Motor Neuron Diseases

UMN onset

ALS

LMN onset

PLS

PMA

ALS plus syndrome

PLS Continued

• Typically affects lower extremities first, then may progress to upper extremities (bottom-up).
• Spastic dysarthria eventually develops, but may take many years to present.
• Dysphagia may be present, but is often less debilitating than with other motor neuron diseases.
• Less commonly begins with bulbar involvement (top-down).
**PLS Epidemiology**

- Prevalence – 1 in 400,000.
- Mean age of onset is between 45-53, wide range.
- Slight male predominance (similar to ALS).
  [Singer et al., 2007]
- Favorable prognosis compared to other MNDs – years to decades.

**Progressive Bulbar Palsy (PBP)**

- Motor neuron disease that may progress into Bulbar-onset ALS.
- Degeneration of the UMN and LMN supplying **ONLY** the bulbar muscles (i.e., tongue, palate, pharynx, larynx, face, jaw).
- Spinal muscles are not affected.
- Dominant features: progressively worsening dysarthria and dysphagia.

**PBP Continued**

- Positive bulbar signs (dysarthria & dysphagia).
- Negative motor dysfunction of the trunk, limbs or eyes. No disturbance in walking, limb and eye movements.
- Negative sensory deficits.
  [Goozée, J. V. & Murdoch, B. E., 2009]

**PBP Epidemiology**

- Incidence - Rare.
- Unlike ALS or PLS, PBP affects women more than men.
- More common in older adults.
- Poorest prognosis of the MND subtypes with death typically <3 years post onset.
  [Goozée, J. V. & Murdoch, B. E., 2009]
- Death occurs due to respiratory failure and swallowing dysfunction.

**Pseudobulbar Palsy**

- Similar to Progressive Bulbar Palsy **EXCEPT** only affects UMN supplying bulbar muscles.
- No LMN signs, such as fasciculations or atrophy.
- No spinal muscle involvement.
- Progressive spastic dysarthria and dysphagia.
- Pseudobulbar affect may be present.
- Expressionless or “masked” face.
- May involve frontal lobe involvement – cognitive executive function impairments.
  [Miller, R. & Britton, D., 2009]

**Progressive Muscular Atrophy (PMA)**

- Motor neuron disease that may progress into Spinal-onset ALS.
- Pure LMN disease that starts as focal spreading to upper and lower extremities, bulbar and respiratory musculature.
- No UMN signs, such as spasticity or hyperreflexia.
PMA Continued

- Slowly progressive muscle weakness.
- Muscle atrophy and loss of muscle bulk resulting in weight loss.
- Fasciculations.
- Fatigue.
- Cramps, aching muscles.

Amyotrophic Lateral Sclerosis (ALS)

“A progressive, degenerative motor neuron disease of unknown cause. Muscle atrophy and spasticity in limb and bulbar muscles result in weakness and loss of ambulation, oropharyngeal dysfunction, weight loss, and ultimately respiratory failure.” (Miller, et al., 1999, p. 1)

ALS

UMN and LMN signs/symptoms can be present in the 4 central nervous system regions:

1) Brainstem → Bulbar Muscles
2) Cervical
3) Thoracic
4) Lumbrosacral

El Escorial World Federation of Neurology Criteria for the Diagnosis of ALS

<table>
<thead>
<tr>
<th>ALS Diagnostic Category</th>
<th>Requirements</th>
</tr>
</thead>
<tbody>
<tr>
<td>Definite ALS</td>
<td>LMN and UMN signs in 3 regions of the body</td>
</tr>
<tr>
<td>Definite familial ALS</td>
<td>LMN and UMN signs in 1 region of the body plus laboratory-supported identification of gene mutation associated with ALS</td>
</tr>
<tr>
<td>Probable ALS</td>
<td>LMN and UMN signs in 2 regions of the body (some UMN signs rostral to LMN signs)</td>
</tr>
<tr>
<td>Probable ALS (laboratory supported)</td>
<td>LMN and UMN signs in 1 region of the body plus electromyographic evidence of acute denervation in 2 or more muscles in 2 or more limbs</td>
</tr>
<tr>
<td>Possible ALS</td>
<td>LMN and UMN signs in 1 region of the body</td>
</tr>
</tbody>
</table>

PMA Epidemiology

- Incidence – Affects 5-7% of all people living with MND. (http://www.mndassociation.org/life_with_MND/pmapls/what_is_PMA.html)
- Men are affected more than women.
- Average Age of Onset – Under 50 years.
- Rate of progression is slower than ALS.
- Mean survival of 159 months (13.25 years) in one study of 17 cases. (Norris, F., et al., 1993)
- 3 year survival of 61.3% in another study of 155 cases. (Chio, A., et al., 1985)
### Motor Neuron Damage Revisited

**Upper Motor Neurons**
- Spasticity or Increased Tone
- Weakness (mild-moderate)
- Clumsiness or Loss of Dexterity
- Hyperreflexia
- Pathological Reflexes
- Pseudobulbar Affect (Emotional Lability)

**Lower Motor Neurons**
- Weakness of Skeletal & Bulbar Muscles (moderate – severe)
- Hypotonia
- Hyporeflexia
- Fasciculations
- Muscle Atrophy or Wasting
- Cramps

### Bulbar-Onset ALS
- Approximately 25% of all cases present with bulbar-onset form with dysarthria and dysphagia as the initial problem. ([Murray, B. & Mitsumoto, H., 2006](#))
- The tongue is typically the first bulbar muscle to show signs of involvement.
- Faster progression of the disease than Spinal-Onset ALS.
- Bulbar onset is associated with shorter survival. ([del Aguila, 2003](#))

### Spinal-Onset ALS
- More common than bulbar-onset.
- Onset in limbs, trunk and muscles of respiration rather than bulbar musculature.
- Slower progression to bulbar musculature than Bulbar-Onset ALS progresses to limb musculature.
- Longer survival than bulbar-onset ALS.

### Familial ALS
- Familial or Inherited ALS = 10% of all diagnosed cases such that individuals have at least one affected family member. ([Jackson, C.E. & Bryan, W.W., 1998 and Siddique, T. & Dellefave, L., 2006](#))
- Average age of onset is about a decade earlier than sporadic ALS. ([Siddique, T. & Dellefave, L., 2006](#))
- Spinal onset is more likely than bulbar onset in Familial ALS.

### ALS-Plus Syndrome
- Meets clinical criteria for ALS but also include 1 or more additional features such as dementia, geographic clustering, extrapyramidal signs, objective sensory loss, autonomic dysfunction, cerebellar degeneration, or ocular motility disturbance.
- We will discuss frontotemporal dementia in ALS later in this presentation.

### ALS Prognostic Indicators
- **Age:** Mean disease duration in sporadic ALS was 72 mos. for those under age 45 and was 33 mos. for those over 55. ([Naric, et al., 1993](#))
- **Gender:** Most studies have NOT shown a survival difference between males and females ([Murray, B., 2006](#))
- **Malnutrition:** (low body mass) increased the risk of death by 7.4 times. ([Desport, J.C., et al., 1999](#))
- **Occurrence of malnutrition was equal in both spinal- and bulbar-onset ALS.**
**ALS Prognostic Indicators**

- Psychosocial: When age, disease severity and duration were controlled, the relative risk of death in patients with psychological distress was increased 2.24 times compared to patients with psychological well being. (McDonald, E.R., et al., 1994)

- Specialist Care: In a study by Chio (2006), mean survival was longer in specialized ALS clinics (1080 days or 2.96 years) than in general neurology clinics (775 days or 2.12 years).

- Multidisciplinary care: An independent predictor of survival and reduced the risk of death by 47% in a 5 year study by Traynor in 2003.

**ALS Prognostic Indicators: El Escorial Criteria**

<table>
<thead>
<tr>
<th>El Escorial Criteria at Diagnosis</th>
<th>Mean Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Definite ALS</td>
<td>23 Months</td>
</tr>
<tr>
<td>Probable ALS</td>
<td>34 Months</td>
</tr>
<tr>
<td>Possible ALS</td>
<td>35 Months</td>
</tr>
<tr>
<td>Suspected ALS</td>
<td>58 Months</td>
</tr>
</tbody>
</table>

Chio, et al. (2002) (Definite ALS vs. Other categories, p < 0.0001)

**ALS-FRSr Predicting Survival Time**

- The risk of death or tracheostomy progressively increased from highest to lowest quartile.
- The Respiratory subscore ($p < 0.001$) and the Gross Motor subscore ($p = 0.02$) were independent significant predictors of death/trach.
- Respiratory subscore was strongest – death in ALS is ultimately due to respiratory failure.

**ALS-FRSr Predicting Survival Time**

<table>
<thead>
<tr>
<th>Total Initial ALS-FRSr Score</th>
<th>Hazard Ratio (CI 95%)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>≥43</td>
<td>1.00 (Reference)</td>
<td>-</td>
</tr>
<tr>
<td>≥38 to &lt;43</td>
<td>7.08 (2.12 – 23.66)</td>
<td>0.001</td>
</tr>
<tr>
<td>≥33 to &lt;38</td>
<td>12.51 (3.81 – 41.08)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>&lt;33</td>
<td>27.78 (8.57 – 90.07)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Kaufman, P. et al. (2005)

**Terminal Stage of ALS**

- Most patients with ALS die of progressive respiratory failure.
- The majority of patients with ALS die within 3-5 years of diagnosis.
- Respiratory status is an important predictor of survival duration.
- Palliative care and hospice care are usually involved in the pre-terminal and terminal stages of the disease progression.

Kaufman, P. et al. (2005)
### COMMUNICATION IN ALS
Kim Winter, M.A., CCC-SLP

"If all my possessions were taken from me with one exception, I would choose to keep the power of communication, for by it I would soon regain all the rest."
Daniel Webster

### Dysarthria and ALS
- “25%-30% of ALS patients have dysarthria as a first or predominant sign in the early stage of the disease.”
  - (Tomik, B. & Guiloff, R., 2010, p. 5)
- “On average, the diagnosis of ALS is made approximately 6 months after the appearance of initial symptoms.”
  - (Ball, L. et al., 2002, p. 232)
- UMN and LMN decline results in a mixed (spastic and flaccid) dysarthria.
- Towards the end stages of the disease, flaccidity predominates.

### Flaccid versus Spastic Dysarthria

**Flaccid (LMN)**
- Hypnasality
- Imprecise consonants
- Breathiness
- Monopitch
- Nasal emission
- Audible inspiration
- Harsh vocal quality
- Short phrases
- Monoloudness
- Lingual atrophy
- Lingual fasciculations

**Spastic (UMN)**
- Imprecise consonants
- Monopitch
- Reduced stress
- Harshness
- Monoloudness
- Low pitch
- Slow rate
- Hypernasality
- Strained-strangled voice
- Short phrases
- Distorted vowels
  - (as cited in Duffy, J., 2005)

### Mixed Dysarthria (UMN & LMN)
- Imprecise consonants
- Hypernasality
- Harshness
- Slow rate
- Monopitch
- Short phrases
- Distorted vowels
- Low pitch
- Monoloudness
- Prolonged phonemes
- Breathness
- Audible inspiration
- Nasal emission
- Reduced stress
- Prolonged intervals
- Inappropriate silences
- Strained-strangled voice
- Excess and equal stress
  - (as cited in Duffy, J., 2005)

### Compensation Not Remediation

“Although advances in understanding the pathophysiology of ALS have stimulated the development of new drug therapies, the mainstay of treatment for ALS patients remains symptomatic management.”
  - (Miller, R.G. et al., 1999, p. 2)

“As with any incurable disease, the state of the art in treatment for ALS is symptom management (also referred to as ‘palliative care”).”
  - (Mathy, P., n.d., para. 9)

### Compensation/Management versus Remediation/Treatment

**Compensation**
- Focus is on speech comprehensibility.
- Energy conservation techniques are utilized.
- There is an increased need for compensatory strategies to supplement natural speech with disease progression.
- Recognition of communication variables.

**Remediation**
- Focus is on speech intelligibility.
- Strengthening activities (i.e. oral motor exercises) and speech drills are utilized.
- Compensatory strategies are gradually withdrawn as natural speech improves (restored function is the anticipated outcome).
- Afflicted individual is the primary consideration.
Speech Intelligibility

- “The degree to which the acoustic signal (the utterance produced by the dysarthric speaker) is understood by a listener”. (Yorkston, K. et. al., 1996, p. 55)
- Typically measured by having listeners orthographically transcribe words or sentences that the speaker reads.
- Has received the most focus in dysarthria research.

Speech Comprehensibility

- “Provides an estimate of the adequacy of performance in natural communication settings”. (Yorkston, K. et. al., 1996, p. 56)
- “Incorporates signal-independent information such as syntax, semantics, and physical context”. (Yorkston, K. et. al., 1996, p. 55)
- Communication partners play a pivotal role.

Energy Conservation: Contraindications of Oral Motor Exercises and Speech Drills

- Lack of empirical evidence to support oral motor exercises (OMEs) in motor speech remediation.
- McCauley, R. et al. (2009) literature review:
  “...the existing research literature provides insufficient evidence to support or refute the use of nonspeech OMEs”. (p. 353)
  “At this time, based on theory and available evidence, the use of OMEs must be considered exploratory, and clients should be informed of this prior to initiating their use in treatment”. (p. 356)

Contraindications of Oral Motor Exercises and Speech Drills Continued

- Bandaid on a gunshot wound approach – “Because of the pathophysiology and the degenerative nature of ALS, speech treatment strategies that are designed to increase strength or mobility of the oral musculature are NOT recommended”. (Ball, L. et. al., 2007, p.290)
- Implications of “false hope”.
- “…speech exercises emphasizing optimum performance can only prove to be a discouraging reminder of increasing loss of ability”. (Mathy, P. n.d., para. 11)
- Fatigues muscles using non-purposeful tasks.
- Unclear if exercise to fatigue may actually hasten neurologic deterioration.
### Speech Staging System for ALS

- **Stage 1:** No detectable speech disorder.
- **Stage 2:** Obvious speech disorder with intelligible speech.
- **Stage 3:** Reduction in speech intelligibility.
- **Stage 4:** Natural speech supplemented with AAC.
- **Stage 5:** No useful natural speech.

(Yorkston, Beukelman, Strand & Bell [1999])

### ALS Severity Scale

- Developed by Hillel et al., 1989.
- 10 point scale addressing 4 areas: speech; swallowing; lower extremity and walking; and upper extremity dressing and hygiene.
- 10 = Normal; 1 = Non functional
- 200 consecutive patient visits – “for 71% of these individuals, swallowing and speech scores were within 1 point of one another”.

(Yorkston, K. et al., 2004, p. 51)

### What is the SLP’s role at each stage?

#### Stage 1: No detectable speech disorder.
- Patient and family education regarding motor speech changes.
- Discussion regarding voice banking.

#### Stage 2: Obvious speech disorder with intelligible speech.
- Discuss compensatory speech comprehensibility strategies.
- Emphasize the communication partners’ role in communication.
- Discuss energy conservation with regard to speech.

### Stage 3: Reduction in speech intelligibility.
- Continue to encourage use of compensatory strategies, emphasizing need to utilize multiple strategies.
- Initiate AAC evaluation when speech rate is approximately 100-125 wpm. Why???
- “Controversial” options:
  - Voice amplifier
  - Palatal lift

### Supplemented Speech Strategies

- “Speech supplementation is a group of several different strategies that augment the speaker’s natural speech by providing additional contextual information to convey the spoken message”.

(Yorkston, K. & Beukelman, D. [2004])
**Supplemented Speech Interventions**

- **Alphabet Supplementation** – speaker indicates the first letter of each word as they say the word.
- **Benefits** –
  - Reduces speaking rate
  - Allows for increased processing time for the listener
- **Limitations** –
  - Speaker must be able to have adequate upper extremity functioning in order to point/select letters
  - Increases the physical and cognitive demands (speech and pointing).

**Mean percent intelligibility across speakers by cue condition.**

(Hustad, K. et al., 2003, p. 469)

**Supplemented Speech Interventions Cont’d.**

- **Topic Supplementation** – speaker indicates the topic of the message (“I want to talk about the weather”). Particularly useful when the speaker is changing topics of conversation.
- **Benefits** –
  - Provides greater contextual support for the listener by constraining listener expectations for presented messages.
- **Limitations** –
  - Research demonstrates that it is not as effective as alphabet supplementation.

**Supplemented Speech Interventions Cont’d.**

- **Gestural Supplementation** – gestures accompany or illustrate speech. This can also include pointing to environmental props (i.e. signs).
- **Benefits** –
  - No additional equipment, portable, always available.
  - Idiosyncratic.
  - Not all messages are “gesturable”.
- **Limitations** –
  - Impairments in upper extremity functioning are a limiting factor.

**Gestures and Alphabet Cues vs. No Cues on Speech Intelligibility**

(Hustad et al., 2005, p. 1004)

**Supplemented Speech Interventions Summary**

- Overall, dysarthric speakers’ speech intelligibility improved when supplemented speech strategies were used.
- Alphabet supplementation and gestures yielded more significant results than topic cues.
- The greater the speech severity, the greater the benefits; however, also more variability in performance. (Hanson, E. et al., 2004)
Stage 3: Reduction in speech intelligibility.
- Continue to encourage use of compensatory strategies, emphasizing the need to utilize speech supplementation strategies.
- Initiate AAC evaluation when speech rate is approximately 100-125 wpm. Why???
- “Controversial” options:
  - Voice amplifier
  - Palatal lift

Speech Intelligibility and Speaking Rate
- Speaking rate is an important predictor of speech intelligibility.
  (Yorkston, K., et al., 1993; Ball, L., et al., 2002; Ball, L., et al., 2005)
- Speaking rate tends to decline before reductions in intelligibility of speech are noted:
  - Is the reduced speaking rate a compensatory strategy (conscious or unconscious)?
  - Is it just an artifact of the patient’s decline in neuro-motor functioning? (Ball, L., et al., 2002)

Relationship Between Intelligibility & Speaking Rate
(Ball, L. et al., 2002, p.234)

Perceived Effort and Speech Intelligibility
(Wilkinson, C. et al., 1995, p.142)

Speech Deterioration: Intelligibility & Months Post-Dx.
(Ball, L. et al., 2002, p.233)

Reasons for a Late AAC Assessment
Assessment was delayed due to...

- Proposal Acceptance: 7%
- Unaware of Services: 4%
- Language Barrier: 7%
- Travel: 13%
- Family Issues: 18%
- Other Health Impairment: 34%
- Self/Group Home: 7%
- Clinic Neurologist: 4%
- Non-Clinic Neurologist: 29%
- General Practitioner: 32%

(Percentage of Late AAC Assessments
(Modified from Nordness, A.S., et al., 2010, p. 52)
Timeliness of AAC Evaluations - “Reality Check”

- At 125 wpm, speech intelligibility is often &gt;=90%. Insurance denial is an issue.
- Early introduction of AAC and patient acceptance: “...early introduction can be a challenging process since it may be quite upsetting for the person who does not yet require AAC to face the reality that this will eventually be the case”. (Doyle, M. & Phillips, B. 2001, p. 169)

Psychosocial Issues and AAC Acceptance/Use

- Partner acceptance and expectations:
  - Will they be responsible for maintenance and programming?
  - Do they have realistic expectations?
- Communication Environment:
  - 1:1 versus group settings; familiar versus unfamiliar communication partners
  - Time of day – fatigue issues

Timeliness of AAC Evaluations - “Reality Check” Cont’d.

- “The key management issue is frequently not device selection, access/interface or vocabulary selection but forced adaptation to altered communication style, loss of spontaneity and potential loss of control”. (Carroll-Thomas, S. 1995, p. 282)

Psychosocial Issues and AAC Acceptance/Use Cont’d.

- Technology skills & age:
  - Are they technology “savvy”?  
  - “Anyone younger than age 30 in 2000 who eventually acquires a communication disorder may respond more favorably to technology-based AAC systems”. (Lasker, J. & Bedrosian, J., 2000, p.115).
- Individual personality
  - Respect patient desires to NOT use AAC.

Stage 3: Reduction in speech intelligibility.

- Continue to encourage use of compensatory strategies, emphasizing the need to utilize speech supplementation strategies.
- Initiate AAC evaluation when speech rate is approximately 100-125 wpm. Why???
- “Controversial” options:
  - Voice amplifier
  - Palatal lift

Psychosocial Issues and AAC Acceptance/Use Cont’d.

- “In clinical practice, some patients choose to communicate to the few close persons who can understand their severely dysarthric speech, or to use the cheapest communication aid (i.e. writing on a piece of paper, alphabet chart). There are also patients who do not use the communication support provided and prefer to remain mute. Their wishes should be respected”. (Tomik, B. & Gukoff, R., 2010, p. 9)
Voice Amplification

• Benefits –
  – Easy to use: no significant training needed
  – Inexpensive: prices average around $300-$400
  – Portable
  – Supports use of natural speech

• Limitations -
  – Hypophonia is often not the only issue
  – Amplifies ALL aspects of the user’s speech, including hoarseness, breathiness, hypernasality, strained/strangled voice, etc.

Palatal Lift Efficacy

Esposito, Mitumoto & Shanks (2000):
• 21 out of 25 (84%) demonstrated reduced hypernasality
• 19 (76%) had moderate benefits for 6 months.
• Patients indicated it was easier to speak with less effort.
• Once severe labial and lingual weakness were observed, palatal lift was no longer beneficial.

The “Reality” of Palatal Lifts

• Fabrication of the prosthesis generally takes 3 sessions – impressions, palatal lift fabrication and fitting/training.
• Hypernasality is not the only issue.
• Progressive nature of ALS.
• Logistical constraints - viewed as “another appointment”.

Who might be a good candidate for a palatal lift?

• Yorkston, Miller and Strand (2004) use the following guidelines:
  1. “Poor velopharyngeal function in the presence of relatively preserved lip and tongue movement
  2. Preserved ability to swallow saliva
  3. Adequate dentition to support the prosthesis
  4. A relatively slow progression of the disorder, suggesting that the person will continue to rely on natural speech as the primary mode of communication for at least several months”. (p. 43)

Stage 4: Natural speech supplemented with AAC

• Continue to encourage use of compensatory strategies to supplement natural speech attempts.
• Procure SGD (Speech Generating Device) equipment and provide training.
• Respect patient desires to NOT use AAC.
Stage 4: Natural speech supplemented with AAC cont’d.

- “In clinical practice, some patients choose to communicate to the few close persons who can understand their severely dysarthric speech, or to use the cheapest communication aid (i.e. writing on a piece of paper, alphabet chart). There are also patients who do not use the communication support provided and prefer to remain mute. Their wishes should be respected.”

(Tomik, B. & Guillef, R., 2010, p. 9)

Stage 5: No useful natural speech.

- Ongoing support for AAC communication (low-tech and high-tech).
- May need to modify existing systems as physical abilities decline.

AAC Services

- What does ASHA say about SLPs and AAC service delivery?
  - 2002 Omnibus Survey: 1,188 SLPs
  - 45% regularly provide AAC
  - Highest percentage of AAC services in hospitals
  - Non-residential health care
  - Residential health care

(ASHA 2004)

AAC Methods

- No Tech: gestures, pointing, talking slowly, exaggerated speech movements.
- Light Tech/Low Tech: eye gaze boards, writing, laser light pointers on an alphabet or communication board.
  - No Tech and Light/Low Tech methods are also considered “Unaided Communication Systems”.
- High Tech: computers and speech generating devices (SGDs).
  - High Tech systems would also be considered “Aided Communication systems”.

What types of AAC are used?

(Mathy, P. et al., 2000)

Bulbar Onset

- Facial expressions
- Gestures
- Respond to yes/no questions
- Handwriting
- High technology SGD

Spinal Onset

- Facial expressions
- Respond to yes/no questions
- Partner dependent scanning
- High technology SGD

Top 5 Median Frequency of AAC Use as Reported by Caregivers (n=34)

Adapted from Fried-Oken, M. et al., 2006, p. 214
AAC Assessment
• “There is no standardized battery of tests that comprise an AAC evaluation....”

ASHA 2004.

HFSC AAC Evaluation
Part 1 - Complete an assessment of:
• Language - receptive and expressive
• Cognition – memory, executive functioning
• Speech/Voice – oral mechanism exam, speech intelligibility

Language and ALS
• Generally intact – screening tools are utilized.
• “Economy of Wording” – Wilkinson, et al., (1995) demonstrated that individuals with ALS often use:
  – Fewer words
  – Shorter sentences
  – More incomplete phrases

Frontotemporal Dementia and ALS
• “A neurodegenerative disorder causing atrophy of the frontal and anterior temporal lobes that can lead to disturbances in behavior and language”. (Wheaton, M., et al. 2007, p.1411)
• Features include: reduced insight, mental inflexibility, distractibility, apathy, problem solving deficits, changes in interpersonal conduct.
• Co-occurrence of FTD and ALS is approximately 15%-20%. (Ringholz, G. & Greene, S. 2006)

Cognition and ALS
(Ringholz, G., et al. 2005)
• 50% of patients had cognitive impairments
  – 30% were mild
  – 20% were classified as having dementia with the majority of subjects being classified as having Frontotemporal Dementia (FTD).
• Deficits were noted in:
  – Executive function, attention, concentration, working memory, verbal fluency, confrontation naming.

Cognition and ALS Cont’d.
(Flaherty-Craig, C., et al., 2006)
• 20 minute standardized screening tools
• Controlled Oral Word Association Test (COWA)
• Neurobehavioral Cognitive Status Examination (COGNISTAT)
• >50% of the non-bulbar ALS subjects and >67% of the bulbar subjects had deficits in at least 1 of 3 measures (verbal fluency, abstract reasoning, judgment), as compared with normal controls.
• Judged to be comparably sensitive to identify deficits as Ringholz et al. study.
Cognition and ALS Cont’d.
• Established an association between sleep disturbance and nocturnal hypoventilation and cognitive dysfunction (memory and executive function deficits).
• With Non-Invasive Positive Pressure Ventilation (Bi-PAP), improvements noted over a 6-week period.
• Respiratory deficits may exacerbate cognitive dysfunction.

Limitations of Cognitive Testing in ALS
• Time limitations – both SLP and patient.
• Cursory screenings may not be sensitive enough to detect subtle changes, particularly in the early stages.
• Severity of motor and speech impairments limits clinical utility of most conventional assessments.

Motor Speech Evaluation
• Speech Intelligibility Testing:
  • Assessment of Intelligibility of Dysarthric Speech (AIDS): Yorkston, Beukelman & Traynor (1984)
    – Measures speech intelligibility for 50 single words and 20 sentences (6-15 words in length)
  • Sentence Intelligibility Test (SIT): Yorkston, Beukelman & Hakel (1996).
    – SIT is a single disk software package that allows the clinician and researcher to administer, score, and store results of a speech intelligibility measurement task.

HFSC AAC Evaluation Cont’d.
Part 2 – Complete SGD evaluation:
• Medicare requires trials of a minimum of 3 different devices.
• Must consider the patient’s current and future capabilities with regard to access (i.e. motor functioning).
• Original work by Yorkston et al. (1993), with updated considerations by Mathy et al. (2000), can assist with decision making.

AAC Considerations
• Group 1: Adequate speech, adequate hand function
  – Intervention: Monitor communication status; provide information to prepare for future communication needs
• Group 2: Adequate speech, poor hand function
  – Intervention: Alternative writing and keyboard access; provide information to prepare for future communication needs
  – Associated with spinal-onset ALS
  [Ball, L. et al., 2007, p.302-303]

AAC Considerations Cont’d.
• Group 3: Poor speech, adequate hand function, adequate mobility
  – Intervention: Alphabet supplementation; alternative writing; portable direct selection low-tech and high-tech AAC options
  – Associated with bulbar onset ALS
• Group 4: Poor speech, adequate hand function, poor mobility
  – Intervention: Similar to group 3 except that AAC options can be mounted to a wheelchair
  – Associated with spinal onset ALS
AAC Considerations Cont’d.
• Group 5: Poor speech, poor hand function, good mobility
  – Intervention: Alternative access (scanning, head or eye tracking, eye pointing, eye linking); may or may not need AAC to be portable
  – Associated with advanced bulbar onset ALS
• Group 6: Poor speech, poor hand function, poor mobility
  – Intervention: Alternative access (similar to group 5). AAC options do not need to be lightweight because the system can be wheelchair mounted.
  – Associated with end-stage ALS (all onset types)

General Considerations During the AAC Evaluation
• Similar to the “car buying” process – many SGD options with many similarities.

General Considerations During the AAC Evaluation Cont’d
• SLPs do NOT need to know everything about every device – use your vendors to help you.
• SLPs DO need to provide patients/families with information regarding the benefits and limitations of SGD's in light of their ALS diagnosis.

General Considerations During the AAC Evaluation Cont’d.
• "Because most individuals with ALS are adults with intact literacy skills, they benefit most from AAC aids and strategies that provide the ability to generate messages through spelling”.

(Mathy, P., n.d., para. 16)
http://www.asha.org/public/speech/disorders/ALSChallenge.htm

Impact of Mechanical Ventilation on AAC Decision Making
• A small percentage choose this option in our clinic, but some report it is growing.
• Mechanical ventilation will extend their life expectancy and duration of AAC use.
• Progression of physical limitations will require consideration of how to modify the selected SGD given motor control decline.

General Considerations During the AAC Evaluation Cont’d.
• The goal of all AAC is **SNUG**: Spontaneous Novel Utterance Generation
• Limitations of Pre-Programmed Vocabulary
• Limitations of Rate Enhancement Techniques:
  – Word Prediction – saves keystrokes, but doesn’t necessarily increase communication rate
  – Abbreviation Expansion – limited use observed
Specific AAC Considerations

- **Access Method** –
  - Direct selection: pointing, joystick, headtracker, eye gaze
  - Indirect selection: scanning

- Direct selection methods are always preferable over scanning
- Regardless of method, access point needs to be consistent and reliable (>80%)
- Fatigue and progression factors

Eye Gaze Considerations

- How it works: A camera emits an infrared signal that tracks eye movements via retinal reflection.
- Developed for the ALS population.
- Several companies offer eye gaze technology, either as an accessory or as a complete package system.
- Trials of more than 1 system is recommended prior to purchase due to variances in eye gaze accuracy.

Eye Gaze Considerations

- Generally NOT viable for individuals who have:
  - Nystagmus
  - Bifocal glasses, even if progressives
  - Inability to focus in one area for a brief period of time

Funding Considerations

- Funding issues need to be discussed at the outset
  - Medicare will pay the lesser of either 80% of their fee schedule or 80% of the cost of the device.
  - Fee schedule changes from year to year
  - Fee schedule is setup according to “E Codes”

January 2010 E Codes Schedule

[http://aac-rerc.psu.edu/index-23807.php.html](http://aac-rerc.psu.edu/index-23807.php.html)

Funding Considerations

- Medicare will NOT cover an SGD once the individual is in hospice care or is in a SNF
- Medicare requires a “dedicated” SGD
  - Can do an “unlock” after received
- Medicare “5 Year Rule” – Fact & Fiction
  - “Usefulness” not <5 years
  - Will pay if second device is a different E code
Funding Example
Say-it! SAM Communicator
- Good for bulbar onset: lightweight and portable.
- Can only direct select via touchscreen (stylus or finger).
- Synthesized speech.
- E2508
- Cost: $3,895

Funding Example
Dynavox Xpress
- Good for bulbar onset: portable, although somewhat heavier than Say-it! SAM Communicator.
- Multiple access methods: touch enter, touch exit, simple touch & scanning.
- Synthesized speech.
- E2510
- Cost: $7,500

Speech Enhancer
Benefits
- Uses natural speech
- Lightweight, portable
- Auto-programs
- Limited training needed

Limitations
- Limited time for use, particularly with bulbar onset
- Expectations of “magic fix”
- Same E code (E2510) as most other SGD

Funding Considerations Cont’d.
- CT Medicaid (Title 19) – will now pay if in a SNF (bed code #32 or 33)
- VA Benefits – patient must be registered with the VA
- CT Tech Act Project – low interest loans specifically for AT, including AAC
- Bureau of Rehab Services

Common technologies as AAC
- Common technologies such as iPhone, iPad, iTouch, that can be modified for a disabled population.
- Applications/software (shareware) are available for minimal cost.
- Good for bulbar onset ALS.
- Limited ability to modify devices to accommodate physical limitations.

“I’m done with the AAC evaluation, now what?”
- Report writing and funding documents: Several online resources to assist
  - [www.aacfundinghelp.com](http://www.aacfundinghelp.com)
  - [http://www.asah.org/slp/healthcare/sgd/cks.htm](http://www.asah.org/slp/healthcare/sgd/cks.htm)
  - [http://www.aac-rerc.psu.edu/index-38242.php](http://www.aac-rerc.psu.edu/index-38242.php)
  - My name is Sarah Baker & I am a speech language pathologist.
If denied by insurance, may need to help with letter writing for an appeal.
Continue to monitor the patient’s status – assist with obtaining an SGD loan while awaiting purchased device.
Informal Clinical Management
Palovcak, Mancinelli, Elman & McCluskey (2007) described the use of rating scales (ALS-FRS-R, ALS Swallowing Severity scale), cervical auscultation during clinical swallow evaluations and use of FEES and VFSS for assessments in their Philadelphia Clinic.
Management included diet modification and thickened fluids, preparatory sets, bolus size, slow rate, cold temperature fluids, carbonated liquids, supraglottic swallow, super-supraglottic swallow and Mendelsson maneuvers as well as chin tuck.
Modifying the method of intake (straw vs. cup vs. spoon, etc.).
They acknowledged the paucity of research to support their approach.
They never mentioned oral care or energy conservation.

Dysphagia Management In ALS:
Current Research
• No efficacy studies available on instrumental vs. clinical assessment approaches to diagnose and to monitor progression of dysphagia in ALS nor QOL outcomes and symptom management.
• No efficacy studies available on the use of oral or pharyngeal exercises on maintaining duration of swallow function in ALS or whether they are harmful.
• No evidence to support or refute the use of thickened fluids in patients with dysphagia due to ALS.

The AAN Practice Parameters (1999).
• The American Academy of Neurology Practice Parameters in ALS (1999) indicates that a careful history should be conducted to identify dysphagia signs and symptoms. Asking the patient specific questions about their chewing and swallowing can be extremely revealing.
• “There is no single test to detect dysphagia in patients with ALS” (p. 7).

Clinical Dysphagia Evals at HFSC
• Includes a thorough history and chart review.
• Review of medications and relevant side effects.
• Cranial Nerve Exam – anticipate chewing/swallowing issues.
• Respiratory Assessment - RT & MD input, patient complaints.
• Oral Assessment – dentition (including visible decay, plaques), xerostomia, oral tissues, oral secretions, oral hygiene.
• Functional Assessment – Independence in performing oral care and self feeding, mobility, overall strength, conditioning and cognitive/language status.
• Nutrition and Immune System – RD and MD input, recent illness, wounds, infections, immunocompromising meds or conditions, unintended weight loss, hydration status.

Variability in Management

HFSC Approach to Dysphagia - Emphasis on Clinical Assessment
• Informal assessment occurs at Team follow-up appointments every 3 months.
• Clinical dysphagia evaluations are performed when patients and caregivers report:
  Fatigue with eating Increased time to consume meals
  Increased coughing or choking Unintended weight loss
  Complaint of worsening pharyngeal sticking
  Difficulty chewing/orally manipulating food
  Respiratory decline interfering with oral intake
  Complaint of increased work/loss of pleasure with eating
  Increased difficulty with managing oral nutrition and hydration intake

SWALLOWING IN ALS
Jen Chapin, M.S., CCC-SLP
“One of the very nicest things about life is the way we must regularly stop whatever it is we are doing and devote our attention to eating.” Luciano Pavarotti and William Wright, Pavarotti, My Own Story
**Clinical Dysphagia Evals at HFSC**

- Feeding trials – Aspiration is not our primary concern. Adequate oral nutrition and hydration is. What can we offer patients to achieve this?
- Consider energy conservation when offering strategies (Big Picture). Suggesting a patient with ALS perform a chin tuck and double swallows for every bite, while reducing aspiration risk for that bite, is going to cause fatigue and inadequate nutrition and hydration over the course of a meal.
- Consider patient preference and QOL needs.

**MBS/VFSS**

- **Pros:** able to visualize all swallow stages, identify physiologic swallow breakdown, visualize aspiration/residue, cause and effects of maneuvers to reduce it, observe effectiveness of cough response to aspiration.
- **Cons:** exposure to radiation (fluoro > static images), barium can be constipating, cost is high, unlike eating an actual meal, unable to determine fatigue effects, transfer to an MBS chair, a snapshot in time.

**FEES/FEESST**

- **Pros:** Assess the structure and function of the velopharyngeal port, pharynx and larynx, visualize presence of secretions, uses real food, may extend time to simulate a meal, maybe done in wheelchair or at bedside, able to assess fatigue, view pre- and post-swallow physiology.
- **Cons:** Unable to visualize actual moment of swallowing (whiteout), no direct assessment of oral or esophageal phases, tube in the nose, unable to view UES opening, cost is lower than MBS but higher than clinical exam.

**Additional Instrumental Considerations**

- Many patients with ALS feel poked and prodded already with having EMGs, blood work, sometimes MRIs and CT scans and so on.
- Once they know their diagnosis and prognosis, some will not want to be put through more tests.
- Most are looking for ways to improve their quality of life and comfort in the simplest way possible.
- So consider this when you are contemplating an instrumental swallow exam.
- Consider, too, that elimination of aspiration is pretty much impossible. Modify a diet all you want, saliva is still being aspirated.

**Instrumental Evaluations**

- Not objective – high degree in variability in interpretation of MBS and FEES among experienced SLPs.
- When recommending an instrumental evaluation, what clinical questions do you have that you need to answer to determine management of your ALS patient’s dysphagia that you were unable to answer from a thorough clinical evaluation?
- Is the result of an instrumental exam going to change my plan of care with this patient?
- Is the result of an instrumental exam going to change the patient’s functional outcome?

**Patient Choice**

- If I have a concern, a clinical question that I cannot answer from doing a thorough and comprehensive clinical dysphagia evaluation with an ALS patient, I will offer the patient the choice of doing an instrumental exam. Allow them to be a part of their medical decision-making.
- Present the pros and cons and your reasoning for considering an instrumental exam.
- Document your education, their indication of understanding and their decision.
Langmore et. al. (1998)

Predictors of Aspiration Pneumonia: How Important is Dysphagia?

- Prospective study involving 189 elderly patients from acute care, outpatient and extended care facilities followed for 4 years for an outcome of aspiration pneumonia (AP).
- 41 of the 189 patients (21.7%) developed AP, consistent with US prevalence data.

Langmore et. al. (1998)

Factors Significantly Associated with AP

- 81% of patients who developed AP had oral-pharyngeal dysphagia.
- Which means that 19% of those who developed AP did NOT have oral-pharyngeal dysphagia.
- 47% of patients who did not develop AP had oral-pharyngeal dysphagia.
- Of the documented aspirators, only about 38% developed AP.
- 28% of those with AP had GER
- 27% of those with AP were tube fed (NPO).

Langmore et. al. (1998)

Significant predictors of AP were:
- Dependent for oral care
- Tube fed before AP
- Dependent for feeding
- Number of medications
- Smoking
- Multiple medical diagnoses
- Number of decayed teeth

Langmore et. al. (1998)

“All of the factors that directly measured dysphagia were eliminated as significant predictors of AP” (p. 76).

Thus, “dysphagia by itself is not sufficient to cause pneumonia” (p. 76).
Langmore et. al. (1998)

Bacterial flora in the oral cavity and pharynx can be altered by:
- Severe underlying disease
- Inactivity
- Malnutrition
- Presence of oral & dental disease
- Medications

Langmore et. al. (1998)

“The shedding of bacteria from the buccal mucosa, tongue dorsum, gingival sulcus and the teeth is about \(10^{11}\) bacteria per day. Plaque, gingivitis, periodontal disease and tooth decay will alter the flora within the mouth and could change the bacterial composition of saliva. Reduced salivary flow, a common side effect of many medications, increases the concentration of bacteria in saliva, and if saliva is aspirated…”

Langmore et. al. (1998)

“…up to 100,000,000 bacteria/ml saliva could enter the lungs” (p.76).

“Once aspiration has occurred, host defenses must rally to clear the material. Cough and mucociliary clearance act to mechanically drive the material out of the lungs, and lymphatics and alveolar macrophages represent the cellular level of host response” (p. 77).

Langmore et. al. (1998)

“Smoking, COPD, CHF or weak cough secondary to neurologic disease could all impair clearance, as would immunocompromised health status” (p. 77).

Aspiration Pneumonia In ALS –

- 40 ALS cases in Olmsted County, MN (Mayo Clinic) followed between 1990-2005 (15 yrs.)
- 23 males, 17 females. (1.3:1)
- 12 Bulbar-onset, 28 Limb-onset
- Mean survival = 26 mo., Median = 18 mo.
- Aspiration pneumonia occurred in 5/40 patients (13%).
- All with AP had prominent dysphagia and chronic compensated respiratory failure.

Aspiration Pneumonia In ALS –

- PEG was placed in 12 of 40 patients.
- PEG placement more likely in Bulbar-onset patients (8 vs. 4 Limb-onset)
- Mean time for placement for Bulbar-onset was 13 months vs. 17 months for Limb-onset.
- 6/12 underwent PEG placement with FVC >50% of predicted. 4 were <50% predicted. 2 were unknown.
- 2/12 with PEG died within 1 week of PEG. Both had FVC <50%.

- Residence in a nursing home was a significant risk factor for development of aspiration pneumonia.
- ALS patients residing in a nursing home were over 7 times more likely to develop AP.
- There was no apparent association with continued oral intake or with medications.
- AP was not more common in bulbar-onset patients, nor was PEG tube placement protective (3 patients with PEG tubes, 2 patients without).
- Survival of those with AP was not significantly different than those without AP (26 months for both).

**Aspiration Pneumonitis**

- “Acute lung injury characterized by acute inflammation of the lung airways and parenchyma after the inhalation of gastric contents” with radiographic findings very similar to those of aspiration pneumonia (p. 281).
- This makes the diagnosis of aspiration pneumonia quite challenging.
- Aspiration pneumonia dx often over-applied.

(Yoon & Steele, 2007)

**Pathogenesis of Bacterial Respiratory Infection**

Respiratory tract below the larynx is sterile in normal, healthy adults.

Infection occurs with one or more of the following conditions:

- Deficient host defenses
- Highly virulent pathogen
- Overwhelming inoculum (high bacteria load)

(Scannapieco, 1999)

**Pathogenesis of Bacterial Respiratory Infection**

Contamination of lower airway by

1. Inhaled aerosolized droplets
2. Aspiration of oral secretions containing microorganisms

First, oral and pharyngeal surfaces must be colonized by oral and respiratory pathogens which are shed into secretions that then contaminate the pulmonary tree.

(Scannapieco, 1999)

**Bacterial Plaque – Biofilm**

- Covers all the oral structures
- Fundamentally bacteria
- Yellowish-white deposit
- Adheres strongly to teeth
- Cannot be dislodged by chewing or jets of air or water
- Primary colonization lasts 4–24 hours with mainly aerobic bacteria
- Secondary colonization lasts 1–14 days & bacteria rapidly multiply
- Plaque thickness increases with anaerobic microorganisms dominating deeper layers
- ~2 weeks later mature plaque forms
- Mature plaque may mineralize and form calculus

(Puy, 2006)

**The Importance of Saliva**

- Saliva is sterile when it leaves the salivary glands
- 93% by volume is secreted by the major salivary glands (parotid, sublingual and submandibular).
- 7% by minor glands, mainly mucous
- Parotid gland – serous secretion, alpha-amylase
- Submandibular – mucins, calcium
- Sublingual – mucins

(Puy, 2006)
Saliva – More About Spit
• Daily Secretion Rate: 500 – 700 ml
• Average mouth volume: 1.1 ml
• Production controlled by autonomic nervous system
• Resting secretion rate: 0.25 – 0.35 ml/minute, submandibular & sublingual glands
• Stimulated secretion rate as high as 1.5 ml/min.
• Greatest volume before, during & after meals.
• Peak around noon & production falls substantially during sleep. (Puy, 2006)

Functions of Saliva
• Lubrication
• Antimicrobial Action
• Maintaining Mucosal Integrity
• Cleansing
• Acid Buffer Capacity & Remineralization of Teeth
• Preparing Food For Swallowing
• Digestion
• Taste
• Phonation (Puy, 2006)

Hyposalivation
Affects QOL and oral health
• Xerostomia = dry mouth feeling
• Frequent thirst
• Difficulty swallowing
• Difficulty speaking
• Difficulty eating dry foods
• Need to drink water frequently
• Difficulty wearing dentures
• Pain and irritation of oral mucosa
• Burning feeling in the tongue
• Dysgeusia = distortion of taste sensation
(Puy, 2006)

Hyposalivation
Signs to watch for:
• Loss of glossiness of oral mucosa
• Dryness of oral mucosa
• Thin, cracked oral mucosa
• Fissures in the tongue dorsum
• Thick saliva
• Angular cheilitis (sore, reddened, swollen cracks or fissures in the corner of the mouth)
• Increased frequency of oral infection, especially Candida sps.
• Presence of dental caries in atypical locations
• Increased size of major salivary glands
(Puy, 2006)

Xerostomia
Xerostomia
• May increase the risk of bacterial colonization in the oral cavity and pharynx
• Which may increase the risk of pneumonia
• Reduced salivary flow results in poor clearance of oral bacteria
• Increases the potential for aspiration of a much larger bacterial load.
• Makes oral care all that more important! (Limeback, 1998)
Xerostomia

• Common in the elderly, both from normal aging and associated with medication side effects.
• Deepens the fissures in the tongue
• “As the volume of saliva decreases, its washing, antimicrobial and buffering capacity diminishes, the oral pH decreases and the bacterial and fungal load increases” (p. 283). (Yoon & Steele, 2007)

The SLP and Oral Care Connection

Proper oral hygiene influences:
• Swallowing
• Taste Sensation
• Speech Intelligibility
• Social Interactions and Confidence
• Patient Comfort
• Dental Health
• Respiratory Health

Oral Hygiene Recommendations

3 Major Components Shared with Hand-Washing

• Friction – loosen bacteria, plaque and debris
  Toothettes do not provide enough!
• Antiseptic – kill bacteria. Be careful of antibiotics resulting in increasingly resistant strains of bacteria. Avoid alcohol based antiseptics.
• Time – minimum of 2 minutes

Friction

• Soft bristled or electric toothbrush is the best.
• Electric or child-sized may work well for better access.
• Suction toothbrushes available for vent patients or those who have severe dysphagia.
• Toothettes only good for removing debris.

Antiseptic/Antiplaque

• Avoid alcohol – drying to oral mucosa.
• Chlorhexidine – apply to teeth and oral surfaces after brushing and suctioning (ICU, vent patients). Only for short-term use for high-risk patients.
• Cetylpyridium chloride – oral antiseptic and antiplaque solution (Sage products)
• Biotene products work well with xerostomic patients.
• Nystatin – effective for Candidiasis/yeast/thrush
• Hydrogen peroxide – oral debriding solution
• Avoid lemon-glycerin swabs – cariogenic and very acidic.

Oral Hygiene Recommendations

• Daily oral assessment is key in facilities – colonization happens quickly!
• Brush teeth, tongue, palate, inside of cheeks – all oral surfaces to loosen bacteria throughout the oral cavity.
• Minimum of 2x/day.
• Oral hygiene must be performed even if the patient is edentulous!
• Should be performed more frequently for patients who have xerostomia, mouth-breathers or those who are NPO.
Oral Hygiene Recommendations

• For patients with dysphagia, we recommend that they receive oral care PRIOR to PO intake.
• Brush to clean, swab or rinse in between.
• Patients that can perform oral care themselves should be encouraged to do so and assisted as needed.
• Dentures need to be brushed too in order to loosen bacterial plaques. Soaking is not enough.

Xerostomia Considerations

• Associated with Sjogren’s, Parkinson’s, DM, cancer treatment, smoking.
• Very annoying and uncomfortable.
• Avoid toothpastes with sodium lauryl sulphate with xerostomic patients (foaming agent breaks down saliva).
• Avoid alcohol-based oral care products - drying.
• Can be alleviated with water, ice chips, artificial saliva/substitutes and mouth moisturizers, which are available as liquids, gels and sprays. (Most patients I know dislike the gels.)

Oral Hygiene

University of Manitoba - Centre for Community Oral Health
http://www.umanitoba.ca/dentistry/ccoh/ccoh_longTermCare.html#PDF

Sialorrhea & Drooling

Sialorrhea – Excessive Salivation

• True or pure sialorrhea is rare.
• More common in ALS is inability to manage normal salivation due to weakness of the oral and pharyngeal musculature.
• About 50% of patients with ALS experience it.
• Drooling is embarrassing, causes social stigma of the disease, leads to difficult social integration, depression, exacerbates difficulty eating and speaking, as well as perioral chapping.
Treatments for Sialorrhea in ALS

- Tricyclic antidepressants (amitriptyline)
- Atropine sulfate
- Glycopyrrolate
- Hyoscine
- Scopolamine
- Tissues
- Portable suction device
- Botox injections to salivary glands (temporary)
- Irritation of salivary glands (permanent)

(Andrews, J., 2009)

Protocol 201 – Dementia & PD

- Part 1 found that honey thick liquids eliminated aspiration more than nectar, which was also more than thin liquids with chin-down posture during VFSS (all comparisons were significant statistically).
  [Logemann, J. A., et al., 2008]
- Part 2 found that participants drinking nectar thick liquids had a decreased incidence of pneumonia compared to those drinking honey thick liquids over a period of 3 months (significant at $p < 0.10$). All were randomly assigned and performed equally well (no aspiration) or poor (aspirated in all conditions) on VFSS.
  (Robbins, J. et al., 2008)

Robbins et al., 2008 Cumulative Incidence of Pneumonia in Chin Down Posture and Thickened Liquid Groups

Thick Liquid Aspiration - Considerations

- You’ve identified that the patient aspirates with thins. Is their coughing uncomfortable or distressing? Ask the patient if their coughing bothers them. How much are they aspirating? Is it consistently occurring? Do they have a strong cough for airway protection? What’s the condition of their dentition, oral tissues and oral hygiene regimen? Have they had a respiratory infection or pneumonia? Do any postures eliminate the cough? Does changing the method or presentation eliminate the cough?

Dehydration

Mild to moderate dehydration is likely to cause:

- Dry, sticky mouth
- Sleepiness or tiredness
- Thirst
- Decreased urine output — eight hours or more without urination
- Few or no tears when crying
- Muscle weakness
- Headache
- Dizziness or lightheadedness

(http://www.mayoclinic.com/health/dehydration/DS00561/DSECTION=symptoms)
Dehydration

Severe dehydration, a medical emergency, can cause:

- Extreme thirst
- Low blood pressure
- Irritability and confusion
- Reduced speech clarity
- Very dry mouth, skin and mucous membranes
- Little or no urination — any urine that is produced will be dark yellow or amber
- Shriveled and dry skin that lacks elasticity and doesn’t "bounce back" when pinched into a fold
- In the most serious cases, delirium or unconsciousness.

Dehydration Contributes To:

- Constipation
- Fecal Impaction
- UTI
- Cardiovascular Symptoms
- Heatstroke
- Delerium
- Hinders Healing of Pressure Sores
- Reduced Participation in Therapeutic Activities

Increased Risk for Dehydration

- Elderly – Age-related changes predispose.
- Acute Disease Suffers
- Chronic Disease Suffers
- Patients in Residential Facilities
- Semi-Dependent and Dependent Patients
- Patients Who Take in Less Fluid Due to Fear of Incontinent Episodes (May transfer to caregivers, but if it does, it is legally neglect)
- Patients on thickened fluids without an alternate source of hydration.

Dehydration and Thickened Fluids

- No significant difference in absorption between thickened fluids and water,
- BUT patients on thickened fluids have a decreased fluid intake compared to those allowed thin liquids
- Dysphagia patients on thickened liquids were found by Vivanti et al. (2009) to receive most of their fluid from food, rather than beverages.
- Fluid intake is decreased if the beverages are not palatable or if there is a lack of variety.

HFSC Approach to Liquids

- Rarely recommend commercial thickeners; although, we do offer it as a choice for patients who are uncomfortable or distressed by coughing with thin liquids (supercoughers).
- Before that, we suggest naturally thicker beverages like V-8, fruit nectars (Goya), etc. if it makes the patient more comfortable.
- We NEVER thicken water. EVER.
- Be careful about the assumption that if a patient did not aspirate thickened fluids on MBS, that they will not aspirate them during meals.

Swallow Maneuvers – “Chin Tuck”

- Literature describes different head and neck flexion postures. Hard to know what’s what.
- Very clinically popular.
- Physiologically should be used to reduce aspiration before the swallow due to reduced BOT retraction & delayed onset of pharyngeal swallow.
- Variable rates of efficacy at reducing aspiration. Solid studies show it reduces aspiration ≤50% of the time (Ashford, et al., 2009).
- Concern about patients with reduced hyolaryngeal excursion and use of chin to chest posture. Does it further limit anterior movement of the hyoid and laryngeal elevation?
- Probably counterproductive for people with lip weakness.
### “Chin Tuck” Alternative
- Slight chin down posture – “Look at your knees”
- Chin should not drop to chest
- Use gravity to keep liquids in the mouth until patient is ready to swallow in cases of weak posterior tongue with posterior bolus loss.
- Seems less likely to cause pain from deep cervical flexion and interfere with laryngeal excursion
- No empirical evidence to support or refute.

### Other Swallow Maneuvers
- Supraglottic swallow, super-supraglottic swallow, Mendelsohn maneuver.
- Little sound empirical evidence that these techniques work to alter swallow physiology by eliminating aspiration or that they improve swallowing function in patients with neurologically induced dysphagia. (Ashford, et al., 2009)
- Some patients cannot perform them.
- More concerning is the extra effort that patients need to exert to perform these maneuvers. Fatiguing.

### HFSC Approach to Liquids
- If I have to choose between a patient aspirating small amounts of thin liquids without distress vs. thickened fluids with the risk of aspiration, I will choose thins. This is based on my own experience and comfort with a certain level of aspiration.
- Recall that in the Langmore study that 38% of known aspirators developed AP... which means that 62% of known aspirators did not.

### Solids – Oral Prep. & Oral Dysphagia
- Reduce choking risk for patients who are having difficulty with solids due to oral issues.
- Specific food avoidances – patient identified.
- Most patients do this themselves – They know.
- Add extra moisture to aide with chewing and lubrication.
- Dice solids if a patient can still chew, but has difficulty with thorough mastication or fatigue.
- Food process to allow for texture when mastication or tongue lateralization becomes an even greater problem.
- Then puree if necessary.

### Teaching Caregiver Responses to Coughing and Choking
- Teach them the difference.
- Cough is the body’s natural airway defense mechanism, and it’s important.
- Choking is when the airway is obstructed.
- Encourage a cough. Don’t slap on the back. A hand rubbing the patients shoulder is ok. Try not to panic, as it makes the patient panic more.
- Red in the face is ok. Blue in the lips and fingernails is not.
- Heimlich maneuver with choking.

### Energy Conservation Strategies
- Consider smaller, more frequent meals throughout the day rather than 3 large meals.
- Make your meals calorie-dense. Adding extra sauces, gravies and condiments provide moisture and lubrication while adding extra calories, sugar and fat.
- Have a variety of food textures and avoid having several foods that are tough to chew, orally manipulate and swallow at the same meal.
- Consider 1-way straws to save energy.
### Use of Swallowing Exercises

- Easterling and Koster (In Jones & Rosenbek, 2010) assert that an “Active therapeutic swallow exercise regimen is contraindicated, as it promotes muscle fatigue and may promote motor neuron degeneration” (p. 5)
- Clark (2003) notes that fatigue from exercise is rarely discussed in SLP literature.
- Until recovery occurs, muscles cannot produce or sustain forces produced before exercise.
- ALS degeneration may impede recovery.

### Use of Swallowing Exercises

- Clark (2003) – Strengthening exercise in ALS may functionally decrease strength levels without any long-term benefit of increased strength.
- Recommend energy conserving strategies over strengthening exercise programs.
- Applies to use of compensatory strategy selection used during meals as well.

### Use of Swallowing Exercises

- Clark (2005) described published studies looking at gross motor physical exercise in ALS showing some short term benefits.
- HOWEVER, unknown whether similar benefits of specific exercise occurs with ALS patients with swallowing.
- Patients with ALS come to our clinic with varied exercise programs: duration, repetitions, sets, frequency, specific exercises - all different.
- Honestly, we tell them to stop them.

### Considerations – Swallow Exercises and ALS

**Questions to Ask:**
How are you measuring benefit?
How long are patients to continue the program?
What are the criteria to stop the program?
When do you follow-up to determine whether the patient is benefitting or not?
Do you tell patients this may not help and it may make things worse?

### An Example of Tunnel Vision

- FEES protocol: 3-5cc custard (puree), 3-5 cc (milk and water) via straw, cracker (solid).
- Diagnosed pharyngeal dysphagia based on premature spill to vallecula and/or pyriforms, pharyngeal residue after the swallow, penetration or aspiration.
- Defined dysarthria as mild, moderate or severe.

### Tunnel Vision

- They reported a unique advantage of FEES was to assess pharyngeal and laryngeal anatomy and presence of secretions before swallows.
- Yet none of their 17 ALS patients exhibited pooled secretions or abnormal vocal fold mobility.
- 9/17 patients were Bulbar-onset ALS.
- Oral motor assessment was not described in the methods section, but 11/14 patients had tongue ROM WNL, 14/17 had lip closure and facial symmetry (smile/pucker) WNL.
- Yet 11/17 patients were dysarthric.
Tunnel Vision

• Findings were increased depth of bolus flow prior to the swallow requiring thickened fluids (that were not tested during their protocol) and smaller bolus sizes (2-3cc), also not tested.
• Increased bolus retention due to weak pharyngeal swallow requiring smaller bolus sizes and either a small liquid bolus or multiple swallows/bolus to clear.

Who sees the problems here?

Recommendations for Medications

• Avoid pills getting stuck in the pharynx for 2 reasons:
  1) Medication not absorbed into the bloodstream,
  2) Caustic damage to sensitive tissues of the pharynx and larynx resulting in irritation, edema, increased mucus production and possibly permanent scarring.

PEG and J tubes do NOT prevent aspiration pneumonia. Therefore, prevention of AP is NOT an indication for PEG placement in patients with ALS.

Alternate Nutrition and Hydration

• Per the American Academy of Neurology Practice Parameters on ALS (1999), a PEG should be recommended BEFORE the FVC drops to <50% of predicted and not in the preterminal phase of disease progression.
• PEG is indicated with symptomatic dysphagia and weight loss resulting in inadequate caloric intake, dehydration or ending meals early due to fatigue or choking.

Pills

PEG Esophagitis – Prolonged mucosal contact and caustic damage.

Take pills one at a time. Take pills in puree. Take pills in jelly (slick). Halve, crush or take in liquid form. Talk with pharmacist.

Alternate Nutrition and Hydration

PEG and J tubes do NOT prevent aspiration pneumonia. Therefore, prevention of AP is NOT an indication for PEG placement in patients with ALS.
Alternate Nutrition and Hydration

- The AAN Practice Parameters (1999) also indicates that a careful history should be conducted to identify dysphagia signs and symptoms. Asking the patient specific questions about their chewing and swallowing can be extremely revealing.
- “Although a barium swallow study may provide supportive evidence for dysphagia, the indication for PEG in ALS depends on the presence of inadequate oral intake and diminished quality of life due to choking rather than the result of a swallow study” (p. 7).

PEG Tubes Early ALS
AAN Practice Parameters, 1999

PEG Tubes Progressing ALS
AAN Practice Parameters, 1999

Possible Complications of PEG Tubes
In ALS Patients
- Aspiration
- Infection
- Bleeding
- Peritonitis

(Beggs, K., Choi, M. & Travlos, A., 2010)

Aspiration pneumonia is the most common cause of death after PEG placement in general (not just ALS).

(Fox, K.A. et al, 1995 as cited in Plonk, W.M., 2008, ASHA Telephone Seminar)

Case Study – Bulbar-Onset ALS

- 45 year old man (BD)
- Former smoker of 2-3 ppd x 15 years, quit 13 years ago
- Referred to Brain MRI 10/08 – Normal
- Labs – HSV-1 Antibodies
- PCP Diagnosis 11/08 – Bell’s Palsy secondary to acute HSV-1 infection.
- Speech - Exercises for dysarthria.

Case Study Continued

4/09 – Slurring is worse. Left thumb involvement. Complaint of tongue weakness and difficulty eating and drinking.
6/09 – Complaint of lost voice, non-productive cough, SOB, fever, neck cramps, problem with left thumb. PCP – Tonsillitis, Bells Palsy, DJD?
7/09 – Review tests, resolved tonsillitis
7/09 – Neurologist Diagnosis – Medullary CVA that is not visible on imaging tests.
### Case Study

- **7/09** – Bilateral carpal tunnel syndrome with decreased strength in both hands, left greater than right; Reflex testing slightly increased. Slight asymmetry of CN IX – Palate.
- Spine MRI – moderate cervical stenosis C6-C7 and DJD at same level.
- **7/09** – MBS oral and pharyngeal swallow WNL, mild CP hypertrophy.
- **8/09** – Spinal work-up: Atrophy left hand muscles.

### Case Study

- **8/09** – Cervical decompression and fusion C6-C7.
- Re-admitted to acute care day after D/C home – syncope likely due to drug interaction.
- Head CT negative, CXR showed haziness in right base, neck CT showed post surgical changes, neck X-ray showed prevertebral soft tissue swelling at fused level.

### Case Study

- Patient complaint of globus sensation prompted SLP involvement. Per MD D/C summary, he “failed” the first MBS. NG tube placed, which patient reports he pulled.
- He “passed” the second MBS: puree with nectar thick liquids. Pt reports being told his lungs were “filled with food,” however, no diagnosis of pneumonia was documented nor were antibiotics prescribed.

### Case Study

- MD D/C Note: First mention of ALS being included in the differential. Felt it was unlikely given rapid progression.
- Patient had follow-up SLP services per his report. He continued on a modified diet of puree and nectar for 6 months.
- He lost 17 pounds during that period.

### Case Study

- **11/09** – Neurology – Dysarthria, left hand atrophy and weakness persisted. Referred to SLP due to dysphagia post-op.
- SLP evaluated and observed fasciculations and tongue atrophy on clinical exam. Referred the patient to HFSC SLP with ? of motor neuron disease.

### Case Study

- **3/10** – Kim evaluated this patient: mild-mod. Flaccid dysarthria with hypernasality, lingual and labial weakness resulting in imprecise consonant production.
- Clinical dysphagia eval showed mild-mod oral-preparatory and oral dysphagia due to decreased lingual and labial strength/ROM, presence of lingual fasciculations and atrophy resulting in slow mastication. Pharyngeal phase WNL. Referred to Neuromuscular Clinic.
Case Study

• 1 week later – NMC Eval: progressive dysarthria, dysphagia, loss of hand dexterity, cramping in UEs, neck & chest, increased emotionality, tongue atrophy & fasciculations with very slow movements, exaggerated jaw jerk, fasciculations of the UEs, weakness & atrophy of intrinsic hand muscles, intact sensation & coordination.

• Working dx of Progressive Bulbar Palsy, with likely bulbar-onset ALS.

4/10 – EMG showed chronic denervation in 3 spinal areas, which confirmed ALS diagnosis.

Case Study

• 9/10 – SLP Consult in NMC: Progressive moderate mixed (flaccid/spastic) dysarthria characterized by slow rate, consonant imprecision, hypernasality, slightly strained-strangled vocal quality, reduced breath support & speech intelligibility due to lingual, labial & palatal weakness, laryngeal spasticity & respiratory muscle involvement.

• Progressive mild-mod. Oral prep., oral & pharyngeal dysphagia due to labial and lingual weakness & reduced ROM & reduced hyolaryngeal excursion and complicated by fatigue and respiratory compromise with reduced cough force.

Take Home Points

• ALS patients are not all alike in terms of onset, progression, symptoms and prognosis.

• Compensation not remediation.

• Clinical evaluations that include cranial nerve exams are crucial.

• If your exam findings do not match the diagnosis, question it and carefully consider the impact of your education and treatment if the diagnosis is in question.

Take Home Points

If you have a patient with a confirmed or possible Neuromuscular Disease that you are not familiar with or are not sure how to manage, please give us a call or email us!

We are happy to discuss cases with you!

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