Amyotrophic Lateral Sclerosis: Update with a Focus on Diagnosis and Management of Communication Changes
Jeff Searl, Ph.D., CCC–SLP
Department of Communicative Sciences and Disorders
Michigan State University

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Agenda
- ALS as a neurological condition
  - Motor Neuron Disease - Intro
  - A few of the common types
  - ALS
    - Epidemiology
    - Understanding of the etiology & pathogenesis?
    - Signs & Symptoms Presentation
    - Clinical Diagnostic Process
    - Interventions
- ALS a communication disorder
  - Signs & symptoms
  - Communication diagnostic process
  - Interventions

Motor Neuron Disorders
- Group of progressive neurological disorders
  - Abnormality/Destruction of motor neurons
    - Upper Motor Neuron
    - Lower Motor Neuron

Motor Neuron Disorders
- Adults and children
- Men/boys and Women/girls
- Familial and sporadic

Some of the more common MNDs
- PMA
- PBA/PBP
- PLS
- SMA
- ALS
- ALS
- Upper Motor Neuron
- Lower Motor Neuron
- Familial or sporadic
- Genetic
- Environmental
- Viral
- Genetic
- In family members with ALS or FTD
UMN and LMN Signs

<table>
<thead>
<tr>
<th>UMN</th>
<th>Upper Motor Neuron</th>
<th>Lower Motor Neuron</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weakness</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Atrophy</td>
<td>Diverse atrophy</td>
<td>Yes – marked &amp; early</td>
</tr>
<tr>
<td>Fasciculations</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Reflections</td>
<td>Hyper-reflexia</td>
<td>Hypo-reflexia</td>
</tr>
<tr>
<td>Tone</td>
<td>Increased</td>
<td>Decreased</td>
</tr>
<tr>
<td>Nerve Conduction Velocity</td>
<td>Normal</td>
<td>Abnormal (motor nerves: reduced amplitude and delayed onset)</td>
</tr>
<tr>
<td>EMG Denervation</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Distinguishing the MNDs

<table>
<thead>
<tr>
<th>MND Type</th>
<th>UMN Involvement</th>
<th>LMN Involvement</th>
<th>Rate of Progression</th>
<th>Other Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amyotrophic Lateral Sclerosis</td>
<td>Yes</td>
<td>No</td>
<td>Fast</td>
<td>Can be Both</td>
</tr>
<tr>
<td>Primary Lateral Sclerosis</td>
<td>Yes</td>
<td>No</td>
<td>Slow</td>
<td>Can be Both</td>
</tr>
<tr>
<td>Progressive Muscular Atrophy</td>
<td>No</td>
<td>Yes</td>
<td>Slow</td>
<td>Hands usually, then lower body; trunk</td>
</tr>
<tr>
<td>Progressive Bulbar Palsy</td>
<td>No/Some</td>
<td>Yes</td>
<td>Slow/Fast</td>
<td>Largely affects more</td>
</tr>
</tbody>
</table>

UMN: Upper Motor Neuron; LMN: Lower Motor Neuron

Revised El Escorial criteria = No; literature says may have some

Relationships Among

ALS: Epidemiology

- CDC MMWR 11/23/2018
  - USA prevalence 01 to 12/2015 = 5.2 per 100K
  - 16,583 "definite" cases identified
  - 6,250 new "definite" cases 2015
  - Higher prevalence in Midwest likely reflects population demographics (More White)

Substantial Growth in Research Past Decade

- Mostly: genetics and pathogenesis, some drug Tx
- National ALS Registry - 2008
- https://www.cdc.gov/als/
- Culls databases to ID cases
  - Medicare
  - VA Health Admin
  - VA Benefits Admin
- Self-enrollment – web portal
- Biorepository

Understanding of the

- Familial ALS → 5% to 10% cases
- Sporadic ALS genes → environment → time

Classification/Level of Certainty

- Definite → 2 first- or second-degree relatives with ALS (or FTLD)
- Probable → 1 first-degree relative with ALS and gene-positive cosegregation
- Possible → Distinct relation (third-degree or beyond) with ALS
- Sporadic ALS patient with no family history, but positive for a FALS gene
- Sporadic ALS patient with no family history but positive for a FTLD gene
- 2nd degree relation: parents, children, siblings
- 2nd degree relation: grandparents, aunts, uncles

Understanding of the Etiology?

- Less advancement beyond genetic basis
- Assume environmental exposure and genetic risk interplay
- No irrefutable links of environmental factors and ALS

Gene → Time → Environment Model [Al-Chalabi et al., 2013]

ALS: Pathogenesis

- Still largely unknown

ALS – phenotypic heterogeneity

Motor Signs

- Variable UMN/LMN involvement
- Variable site of onset in the body
- Variable rate of progression
- Causative genes not distinctly related to motor phenotype
- But focal onset then progressive spread of motor symptoms → usually along neuroanatomic pathways

ALS phenotype – beyond motor

- Sensory
  - Degeneration and malfunction of sensory neurons in ALS, particularly those with SOD1 mutation (Fregani, et al., 2007; Pradat & El Mendili, 2014)
  - Sensory ‘dying off’ may precede motor deficits in ALS (McElligott, et al., 2009)
  - Distal small-fiber neuropathy (Shefner, et al., 2009; Sullivan, et al., 2007; Sunderland, et al., 1978)

- Behavioral Deficits
  - Anomaly = most commonly reported dimensional Apathy Scale, specific for ALS (Farooqui, et al., 2016)
  - Disinhibition (Tibaldo, et al., 2011)
  - Habitation — more associated with C9orf72 genotype (Nuttall, et al., 2012)
  - Pseudobulbar affect (Beded, et al., 2016)
Relevance to Clinical Care

- Cognitive-Behavioral deficits → Tx adherence, making informed decisions, ability to learn/use assistive devices (Chenius, et al., 2018)
- Cognitive impairment
  - negative prognostic indicator linked to survival (Sahnes, et al., 2015)
  - increased care-giver burden (Andrews, et al., 2017)
  - Reduced QOL (Bock, et al., 2017)
- Inclusion of neuropsychologist as core members of ALS multidisciplinary care teams (Jennett, 2015)

ALS: Prognosis – most recent modeling

- ALS Prognostic Index (API)
  - Validity established in model training (n=117) and testing (n=87) set; then externally validated on another n=122.
  - Risk Group classification
    - Predicted survival time
    - Predicted poor prognosis (<25 month survival from onset)
    - Predicted good prognosis (>50 month survival from onset)

ALS: Diagnostic Process

- Remaining a clinical task – ruling out
  - History
  - Examination
    - UMN/LMN involvement
    - Exclusion of other diseases
    - Imaging
    - Blood work
  - Electromyography and nerve conduction studies often used
  - Muscle biopsy
    - Less often – spinal fluid analysis

Diagnosis of ALS Requirements - AAN

1. Signs of LMN – by clinical, electrophysiological, neuropathological
2. Signs of UMN – by clinical
3. Progression from within a region to other region(s)
4. Absence of electrophysiological evidence of other disease to explain clinical/electrophys signs
5. Absence of neuroimaging evidence of other disease to explain clinical/electrophys signs

- Clinical features required
  - Signs of LMN degeneration in ≥2 body region (bulbar, cervical, thoracic, lumbosacral)
  - Signs of UMN degeneration in ≥2 body region
  - Occurrence of LMN and UMN signs + progression to other region(s) determines certainty of the ALS diagnosis
ALS Multidisciplinary Teams

- Typical Composition
  - Neurologist
  - SLP
  - PT
  - OT
  - Nutrition/Dietetics
  - Respiratory therapy
  - Social Work
  - Genetic Counselor
  - Neuropsychologist

http://www.alsa.org/community/

ALS Multidisciplinary Team Care vs … community care

- Longer survival
- Improved QOL
- Improved access to therapies
- Greater patient satisfaction

Rodriquez et al. (2011). Neurologia, 26; 455‐460
Riemenschneider et al. (2013). Ann Neurol, 65, S24‐S28

Primary Areas of ALS Disease Management

- Disease modifying – pharm
- Symptomatic Treatment
- Palliative Care
  - Nutrition
  - Respiratory
  - Communication
  - ADLs

- Riluzole → 1995 as Tx for ALS
  - Survival benefit ~1 (maybe 6) months
  - No discernable effect on QOL or function

- Edaravone (Radicava®) → 2017 FDA approval
  - IV administration
  - 2wk daily dose → 2 week off
  - Slowly repeated (weekends off) → how many cycles?
  - Can be taken with Riluzole

- High cost – estimated at $145,000/year; VA, many insurance companies limit coverage

ALS Team Quality Measures – SLP related

- Measures and description:
  - ALS multidisciplinary care plan developed or updated
  - Percentage of patients diagnosed with ALS who have a multidisciplinary care plan developed, if one did not previously have one at that time.

- ALS symptom treatment offered
  - Percentage of patients with symptoms that are treated with ALS symptom therapy or palliative care, and ALS medical symptoms.

- ALS physical performance: muscle function testing
  - Percentage of patients with a diagnosis of amyotrophic lateral sclerosis who died before beginning of symptom onset.

ALS Multidisciplinary Team Quality Measures

Primary Areas of ALS Disease Management

- Disease modifying
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  - IV administration
  - 2wk daily dose → 2 week off
  - Slowly repeated (weekends off) → how many cycles?

- Can be taken with Riluzole

- 2001 Japanese approval for stroke → free radical scavenger
  - No discernable effect on QOL or function
  - Generally well tolerated
  - Cost: depends, $500/month no coverage, $20-$200 copays with coverage, Medicare and VA cover

- In ALS application
  - Slowed decline of ALSFRS-R by 33% over 6 months
  - Similarly slowed decline in QOL
  - Respiratory measures trended similarly but not significant

- High cost – estimated at $145,000/year; VA, many insurance companies limit coverage
Primary Areas of ALS Disease Management

- Disease modifying
- Several other ongoing trials
- [https://clinicaltrials.gov/](https://clinicaltrials.gov/)

Symptomatic Treatment

- **Ventilatory support**
  - Assisted ventilation → usually noninvasive (NIV), bilevel positive airway pressure (BIPAP)
  - Preset inspiratory and expiratory pressures delivered via mask
  - Trials:
    - Substantially prolong life [Radunovic, et al., 2017; Burkhardt, et al., 2017]
    - Improve QOL [Birnbaum, et al., 2002]
    - Noninvasive often eventually fails – decision about invasive (trach)

  - AAN → good evidence for NIV to prolong life, slow FVC decline, improve QOL
  - Consider early NIV to improve QOL
  - Speech-related → amplification for use while on NIV

  - **Phrenic Nerve Stimulators** = not effective per RCTs [Gonzalez-Bermejo, et al., 2016; DiPALS Writing Committee, 2015]

Symptomatic Treatment

- **Pseudobulbar affect treatment**
  - Pharmaceutical intervention
    - Dextromethorphan + quinidine → FDA approved
    - Oral med
    - Various side effects possible (diarrhea, stomach pain, cough, dry eyes, muscle spasms, etc.
    - AAN identifies “good” evidence for its use in people with ALS who have pseudobulbar affect
Symptomatic Treatment

- **Pseudobulbar affect**
  - 25%-50% of patients with ALS
  - Nuedexta (DMQ) = dextromethorphan + quinidine
  - Approved 2011 for ALS
  - Interestingly, pt's reported improved speech
  - Center for Neurologic Study – Bulbar Function Scale (CNS-BFS)
  - Speech
  - Swallowing
  - Salivation
  - All domains improved on Nuedexta trial
  - More to come

Nutrition Support

- AAN → if impaired oral intake, consider enteral nutrition with PEG
- Stabilize body weight – good evidence
- Prolong survival – good evidence
- Do not use Creatine – not effective with strong evidence
- Do not use high dose vitamin E – not effective with good evidence

ALS: Speech Features

- Quite variable
  - UMN vs. LMN involvement
  - Specific speech sub-system involvement
  - Other impacts
    - General fatigue
    - Mood, apathy
    - Pseudobulbar affect
    - Ventilatory support needs

Academy of Neurologic Communication Disorders and Sciences (ANCDS)

- Appearance of Speech symptoms?
  - 33 months before Dx – 60 months post Dx

- Type of 1st Speech Symptoms – Varies
  - Laryngeal early in many studies
  - Nasal in some studies
  - Velopharyngeal in some
  - Oral Articulatory precision in some

- Harsh, strain, breathy
  - Articulatory imprecision
  - Hypernasality, nasal emission
  - Slewed speaking rate
  - Increased pause frequency, duration
  - Monopitch
  - Monoloudness
  - Maximum performance – DDK
  - Reduced intelligibility
  - Etc.

- Speech Intelligibility relative to speech changes
  - Physiologic correlates – many
    - Reduced articulator velocity (Turner, 1990)
    - Flattened F2 slope (Turner, 1990)
    - Reduced vowel space (Turner et al., 1990)
    - Speaking rate (Ball et al., 2002)
    - Etc.

- Speech Intelligibility relative to physiologic correlates
  - Reduced articulator velocity
  - Flattened F2 slope
  - Reduced vowel space
  - Speaking rate

- Classically described as mixed flaccid-spastic dysarthria

- Of Note:
  - Speech intelligibility reduction not usually an early bulbar symptom
  - Often with good intelligibility well beyond bulbar symptom presence
  - Once SI decrease starts, often a rapid intelligibility decline
  - Speaking rate at or below 120 wpm predicts decline in SI within a few months
  - Once SI decrease starts, often a rapid intelligibility decline
  - Should prompt AAC referral

- Maximum performance – DDK
  - Reduced intelligibility
  - Etc.
Bulbar Dysfunction Diagnostic – What are people doing?

Authors note: 85% pts have dysphagia → 45% don’t routinely do CSE, 73% don’t routinely do MBS → citing “clinical assessment alone is enough,” “basing Tx decisions on pt report alone,” “immediate PEG if choking or weight loss,” “don’t need it since dysphagia is expected.”

Best Practice – Bulbar Function Diagnosis

• Response to the Plowman et al., 2017 survey results
• Convened working group focused on – clinical speech evaluation, AAC evaluation & swallow evaluation
• Goals:
  1. Standardize bulbar data collection across sites
  2. Develop referral guidelines for speech, AAC, swallow
  3. Establish common data elements for speech, AAC, swallow evaluations

Muscle & Nerve, epub ahead of print

Speech Referral Guidance

1. Initial clinic visit – speech evaluation
2. At all visits – gather this
3. Follow-up visits – speech evaluation is integral component but frequency/duration may vary
   • Patient needs
   • Clinic resources
   • Etc
4. All patients with atypical oral motor exam – Otolaryngology referral

What’s a meaningful ALSFR-R change?

• 90% clinicians (n = 65) indicated a 20% change as meaningful (i.e., 4 point change) [Castrillo-Viguera et al., 2010]

A moment on CNS-BFS

• This group deployed the CNS-BFS in the Nuedexta trial – [Smith et al., 2017]
• It was more sensitive to Tx effect than other bulbar measures (speaking rate, swallow)
• Here they validate the scale

• N=120 at 7 sites; 60 were from Nuedexta trial
• Clinic judgement of
  • normal or abnormal speech;
  • clinical swallow assessments (duration measures)
• Salivation – normal or abnormal
• Patients
  • CNS-BFS
  • VAS for speech, swallow, salivation
• Trained evaluator – ALSFRS-R
CNS-BFS
- 21 questions
- Self-administered
- 3 domains
  - Speech
  - Swallowing
  - Salivation
- 7 questions per domain
- Scaled 1-5 (6 on speech items if unable to speak)
- Score range
  - Low of 21 (no bulbar Sx)
  - High of 112
- CNS-BFS (and ALSFRS-R) highly predictive of clinician Dx
- CNS-BFS stronger correlations than ALSFRS-R and patient VAS with timed reading and swallowing

Back to Provisional Guideline: common elements
- Speech Assessment to include
  - Spontaneous sample
  - Reading passage – either/s
  - Rainbow
  - Bamboo
  - Sequential Motion Rate (“puhtuhkuh”)
  - Max sustained /a/
- Clinician Rating of dysarthria severity
  - 0=normal, 4=severe
- Speaking rate (wpm)
- Identification of speech subsystems involved (respiratory, phonatory, articulatory, resonatory)
- Estimated time = 8-10 min

AAC SubGroup Recommendations
- Early AAC exposure and training emphasized
  - Start before overt bulbar Sx
    - “The AAC evaluation should, therefore, be recommended at the time of diagnosis, regardless of whether speech impairment exists.” (Pattee et al., 2018, p2)
  - Initial clinic visit
    - Introduce concept of AAC, broad definition of AAC
    - Arrange referral for AAC evaluation – should be ongoing, repeated as needed as abilities and needs change

Bamboo walls are getting to be very popular. They are strong, easy to use, and good looking. They provide a good background and create the mood in Japanese gardens. Bamboo is a grass, and is one of the most rapidly growing grasses in the world. Many varieties of bamboo are grown in Asia, although it is also grown in America. Last year we bought a new home and have been working on the flower gardens. In a few more days, we will be done with the bamboo wall in one of our gardens. We have really enjoyed the project. [Yunusova et al., 2016]
Swallowing SubGroup

- SLP Swallow Screen
  - Testing/information in 5 domains
    - Patient-report outcomes
    - Diet intake
    - Pulmonary function and airway defense
    - Bulbar function/broadly
    - Dysphagia/aspiration screen

- All patients should undergo swallow screen ➔ failed screening results in referral for comprehensive dysphagia evaluation ➔ VFSS an important component
  - Define issues
  - Assess strategy effectiveness

Swallowing SubGroup – specific tools recommended

<table>
<thead>
<tr>
<th>Domain</th>
<th>Tool</th>
<th>Source</th>
</tr>
</thead>
<tbody>
<tr>
<td>PR-report measure</td>
<td>Eating Assessment Tool–10</td>
<td>Belafsky et al., 2006</td>
</tr>
<tr>
<td>Diet intake</td>
<td>ALS Severity Scale – Swallowing Subscale</td>
<td>Hillel et al., 1989</td>
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<td>Pulmonary Function/Airway defense</td>
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</table>

EAT-10

<table>
<thead>
<tr>
<th>Scale the appropriate response</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
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<th>7</th>
<th>8</th>
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<th>10</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. My swallowing problem has caused me to lose weight</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>6</td>
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</tr>
<tr>
<td>2. My Swallowing problem interferes with my ability to eat for me</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
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<td>10</td>
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<tr>
<td>3. Swallowing affects my ability to speak</td>
<td>0</td>
<td>1</td>
<td>2</td>
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<td>4. Swallowing affects my ability to swallow</td>
<td>0</td>
<td>1</td>
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<tr>
<td>5. Swallowing affects my ability to chew</td>
<td>0</td>
<td>1</td>
<td>2</td>
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<td>4</td>
<td>5</td>
<td>6</td>
<td>7</td>
<td>8</td>
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<td>10</td>
</tr>
<tr>
<td>6. When I swallow food it is too hot</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>6</td>
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<tr>
<td>7. I am able to eat what I want</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
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<tr>
<td>8. Swallowing is difficult</td>
<td>0</td>
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<td>Total EAT-10</td>
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</tr>
</tbody>
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ALS Severity Scale (ALSSS; Hillel et al., 1989)

1. Exclusion Criteria
   - Patients with poor oral intake
   - Patients with poor oral nutrition

2. Administration
   - swallow testing
   - respiratory assessment

Neuromuscular Disease Swallow Status Scale

Yale Swallow Protocol —Sloggy & Leder, 2014

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</tbody>
</table>

ALS Severity Scale (ALSSS; Hillel et al., 1989)

1. Exclusion Criteria
   - Patients with poor oral intake
   - Patients with poor oral nutrition

2. Administration
   - swallow testing
   - respiratory assessment

Neuromuscular Disease Swallow Status Scale

Yale Swallow Protocol —Sloggy & Leder, 2014

1. Exclusion Criteria
   - Patients with poor oral intake
   - Patients with poor oral nutrition

2. Administration
   - swallow testing
   - respiratory assessment

Swallowing SubGroup – specific tools recommended

<table>
<thead>
<tr>
<th>Domain</th>
<th>Tool</th>
<th>Source</th>
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<tbody>
<tr>
<td>PR-report measure</td>
<td>Eating Assessment Tool–10</td>
<td>Belafsky et al., 2006</td>
</tr>
<tr>
<td>Diet intake</td>
<td>ALS Severity Scale – Swallowing Subscale</td>
<td>Hillel et al., 1989</td>
</tr>
<tr>
<td>Pulmonary Function/Airway defense</td>
<td>Forced Vital Capacity (FVC) Cough Test</td>
<td>Plowman et al., 2016</td>
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<td>Pulmonary Function/Airway defense</td>
<td>Oral Motor Exam</td>
<td>Hirakawa et al., 2017</td>
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<tr>
<td>Bulbar function</td>
<td>Oral Motor Exam</td>
<td>CNS-BFS</td>
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<tr>
<td>Dysphagia – Aspiration Screen</td>
<td>Yale Swallow Protocol</td>
<td>Leder &amp; Sloggy, 2014</td>
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EAT-10

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<tr>
<th>Scale the appropriate response</th>
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<tbody>
<tr>
<td>1. My swallowing problem has caused me to lose weight</td>
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<td>2. My Swallowing problem interferes with my ability to eat for me</td>
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<td>4. Swallowing affects my ability to swallow</td>
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<td>5. Swallowing affects my ability to chew</td>
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3. Results/Rx

**Swallowing SubGroup**

- Education of patient/caregiver → stressed
  - Feeding tube role for nutrition/hydration supplementation
  - Need for good oral hygiene – linking to aspiration pneumonia
  - Various swallow maneuvers as needed
  - Modifications to food textures, other diet modifications
  - Pulmonary hygiene, cough, basic life saving techniques

**Provisional Guidelines – what about cognitive & language?**

- They didn’t specify a tool.
- But other pubs and authors heavily using
  - ALS Cognitive Behavioral Screen (ALS-CBS)
  - ALS Caregiver report of behavior

**ALS Cognitive Behavioral Screen – Cog Section**

- Four subsections
  - Attention
    - 2 & 3 step commands
    - Eye-movement
  - Concentration
    - Digit span reverse order
  - Tracking/Monitoring
    - Months backward
  - Alphabet
    - Letter-number alternation
  - Initiation and Retrieval – verbal fluency
    - Words with letter ‘f’

**ALS Cognitive Behavioral Screen – Behavioral Section**

- 15 items
- Changes since disease onset
- 0-3 score per item
  - No change
  - Small change
  - Medium change
  - Large change

**SLP Intervention**

- Limited (no?) change over the years in terms of approach
- Primarily compensatory in nature
- Very little investigation of SLP intervention effectiveness
General Approach

- Optimize speech for as long as possible, including working with partner and family
- Plan for long term communication beyond useable speech, i.e., AAC – including voice banking, message banking
- Complete or help ensure AAC eval is done and Rx made
- Continue to follow regularly to adjust to the patient’s changing needs

A few items that have been reported or evaluated

- What’s been or is tried?
- Evidence?

Prosthetics – velopharyngeal, palatal lowering

Prosthetics – data regarding ALS?

- Esposito et al. (2000) – case series
  - N=25, all ALS
  - Palatal lift = 25
  - Palatal augmentation = 10
  - Measures:
    - “Intelligibility” – hypernasality, articulation
    - Interview re: benefit
  - 84% = reduced hypernasality
  - 100% = pt/family report of benefit (“easier to speak,” “worth the effort”)
  - Of 10 with palatal augmentation = 60% perceived benefit to articulation

- Decker et al. (2012) – single case
  - ALS
  - Palatal lift
  - “The patient stated that her team of speech therapist, neurologist, otolaryngologist and dentist noticed a better pronunciation with significant decreased hypernasality.” p. 561
  - Also reference to Nasometer with improvement noted

- Watanabe et al. (2012) – single case
  - ALS
  - Several types of prosthesis attempted over 13 months
  - Unclear how they tracked speech
  - Concluded: prosthesis was beneficial but only early on

Prosthetics – data regarding ALS?

- Not much to hang your hat on
Exercise to strengthen articulators? No.

- Dworkin & Hartman (1979) –
  - fairly intense tongue strengthening in single case
  - no improvement

- Watts & Vanryckeghem (2001)
  - Single case – female with ALS
  - LSVT to address voice (without improvement)
  - Oral motor movements and strengthening → intelligibility decline continued

Voice Amplification?

- Often hear that it won’t help
- Will only make unclear speech louder?

- Anecdotal evidence from patients
- Most focus on the reduction in “effort” that might happen with amplifier

Voice Banking

- Voice Banking
- Message Banking

Voice Banking

- ModelTalker – for example
  - Create your synthetic voice
  - Loadable/useable in various apps and speech generating devices (e.g., TherapyBox apps such as predictable)
  - $100

Message Banking

- TobiiDynavox Message Banking
- Patients own voice
- Recordings of phrases, sentences, meaningful sayings wanted for subsequent import into speech generating device or apps

Message Banking app

Voice amplification video Boston Children's Bank
And a different type of Banking - Legacy

- [http://www.recordmenow.org/](http://www.recordmenow.org/)
- Video and/or audio
- To leave a lasting set of messages, stories, etc. for loved ones.

Strategies talked about but not evaluated specific to ALS

**Communication Environment**
- Background noise reduction
- Face-to-face when possible
- Select environments (e.g., quieter restaurant)
- Window up in car if needed
- Determine whether eating and speaking are compatible – avoid the combo if needed

Indicates reported in Murphy (2004) as used by patients and families

**Speech Production**
- Some over enunciation – if it is not fatigue inducing
- Slurring of consonants – avoid if possible
- Monitor phrase length – avoid speaking on residual air
- Many think about slowing down, but most often they are already slow
- Talking louder often just increases fatigue
- Repeat if needed
- Verbal spelling
- Emphasizing key words

**Conversation Strategies & Communication**
- Understand your best times of day → fatigue/energy → plan important conversations accordingly
- Partner assist/interpretation
- Topic cueing
- Facial expression, eye contact, gestures
- Look for indications of understanding or lack thereof → repair strategies
- Low tech assists – pointing, alpha board, writing, various pic boards

Partner Prediction Video

Murphy, 2004 – 15pts/partners; self report of what they tried to facilitate communicatin

**Partner Training**
- Awareness of emotional lability – discuss how to handle (e.g., topic change, rib nudge, nothing)
- Partner prediction – potentially helpful, potentially not
- Don’t talk over
- Talking louder to me doesn’t help
- Ask before you decide to speak for me
- I’ll let you know if I am too frustrated and want to stop a conversation.
- Acknowledge when you don’t understand
Sialorrhea treatment

- Sialorrhea in ALS = not increased saliva production; decreased swallowing of saliva

- Anticholinergic medications – usually tried 1st, various outcomes.
  - Some suggest scopolamine patches [McGeachan et al. 2013] – but various side effects
  - About 33% of ALS patients do not respond to anticholinergics
  - And even for those who have initial response, often not safe or becomes ineffective over time

- Botox – ANN good evidence
  - Parotid and submandibular glands
  - Sialorrhea decreases about 3-7 days post injection
  - Max reduction at 2-4 weeks
  - Typical effect for 3.5 months but quite variable

Radiation to submandibular glands – AAN weak evidence

- Electron based better than photon based
- Various dosing schedules attempted
- 4-6 months benefit reported in some studies
- Comparison to Botox → not enough to draw conclusion regarding superiority of one over the other

All Done!

- Questions & Comments?