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# ALS: A Clinical Population with Unique Communication Management and AAC Needs

**Part 1**


*We Rebuild Lives.*

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**September 15, 2016**

## Disclosure Statement

- I received an honoraria from Speech Pathology.com to produce this presentation.
- I have no other financial or non-financial relationships to disclose.

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## Learner Outcomes

1) describe the neurologic underpinnings of ALS

2) describe the differences between a compensation/management approach versus a treatment/remediation approach to SLP interventions

3) describe the Speech Staging System for ALS, the roles of the SLP and various interventions for each stage

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## 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

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# NEUROANATOMY REVIEW

## What is ALS?

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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.

**Project ALS video

http://www.projectals.org/what-is-als

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# ALS

- Lou Gehrig's Disease
- Progressive
- Degenerative
- Upper and Lower Motor Neuron Disease
- Heterogeneous presentations
- No known cause
- No known cure

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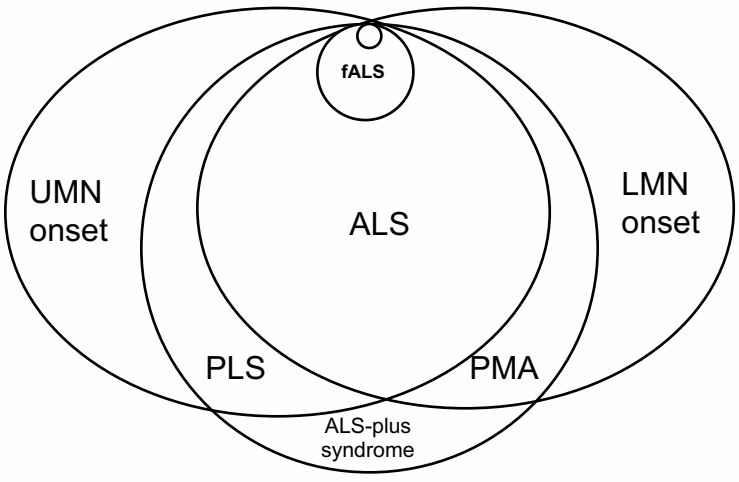
## Spectrum of Motor Neuron Diseases

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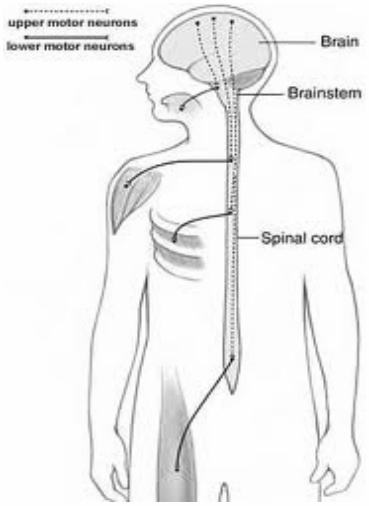
## Review of Neuroanatomy

- 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.

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## Motor Neuron Damage

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

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## Bulbar vs. Spinal Musculature

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

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## 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 shows signs of involvement.
- Faster progression of the disease than Spinal-Onset ALS.
- Bulbar onset is associated with shorter survival.

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## 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.

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## Familial ALS

- Familial or Inherited ALS = 10% of all diagnosed cases such that individuals have at least one affected family member. (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.

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## 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.

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## 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.

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## Flaccid versus Spastic Dysarthria

| Flaccid (LMN) | Spastic (UMN) |
|---|---|
| Hypernasality | Imprecise consonants |
| Imprecise consonants | Monopitch |
| Breathiness | Reduced stress |
| Monopitch | Harshness |
| Nasal emission | Monoloudness |
| Audible inspiration | Low pitch |
| Harsh vocal quality | Slow rate |
| Short phrases | Hypernasality |
| Monoloudness | Strained-strangled voice |
| Lingual atrophy | Short phrases |
| Lingual fasciculations | Distorted vowels |

(Darley, F., Aronson, A. & Brown, J., 1969, as cited in Duffy, J. ,2005)

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## Mixed Dysarthria (UMN & LMN)

| | |
|---|---|
| Imprecise consonants | Prolonged phonemes |
| Hypernasality | Breathiness |
| Harshness | Audible inspiration |
| Slow rate | Nasal emission |
| Monopitch | Reduced stress |
| Short phrases | Prolonged intervals |
| Distorted vowels | Inappropriate silences |
| Low pitch | Strained-strangled voice |
| Monoloudness | Excess and equal stress |

(Darley, F., Aronson, A. & Brown, J., 1969, as cited in Duffy, J. ,2005)

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## 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)
http://www.asha.org/public/speech/disorders/ALSChallenge.htm

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## Compensation/Management versus Remediation/Treatment

| Remediation | Compensation |
|---|---|
| Focus is on speech intelligibility. | Focus is on speech comprehensibility. |
| Strengthening activities (i.e. oral motor exercises) and speech drills are utilized. | Energy conservation techniques are utilized. |
| Compensatory strategies are gradually withdrawn as natural speech improves (restored function is the anticipated outcome). | There is an increased need for compensatory strategies to supplement natural speech with disease progression. |
| Afflicted individual is the primary consideration. | Recognition of communication variables. |

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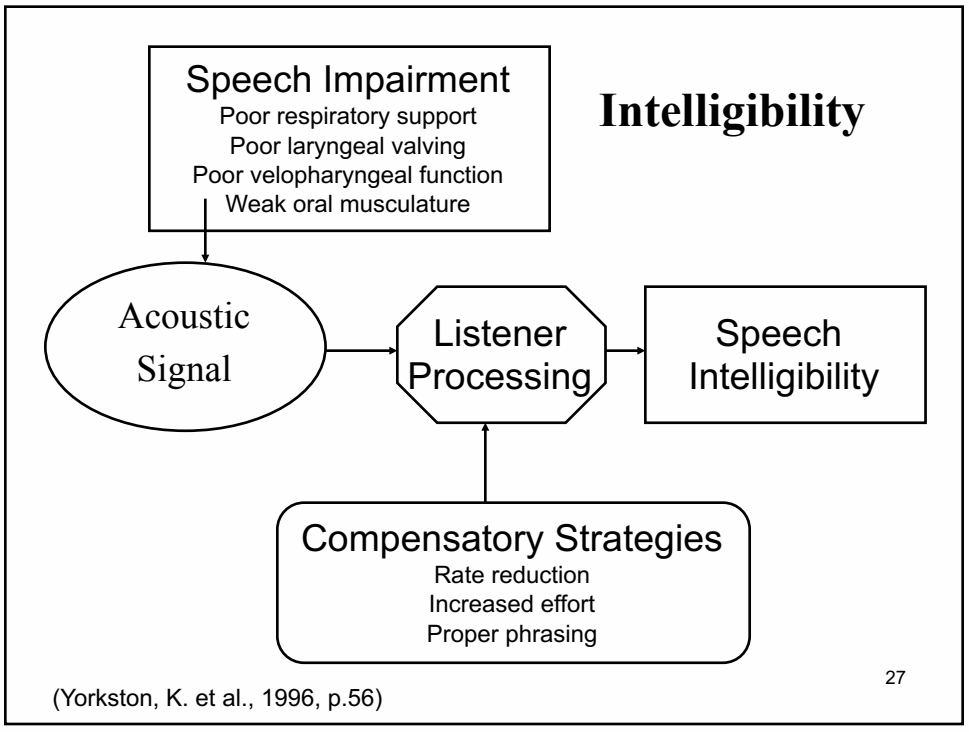
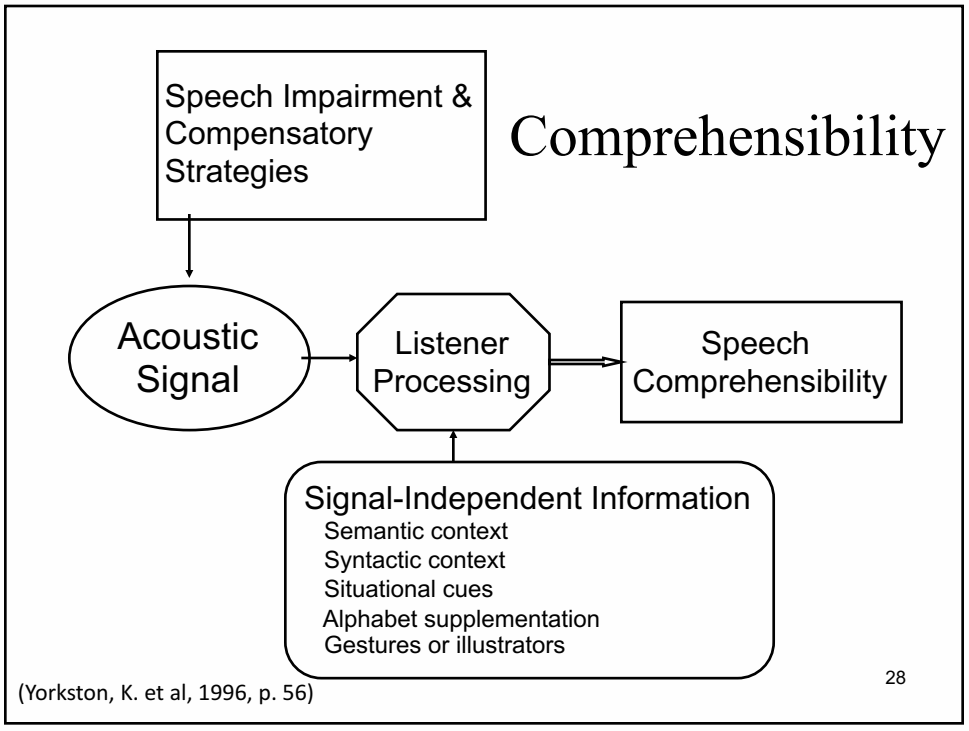



(Yorkston, K. et al., 1996, p.56)




(Yorkston, K. et al, 1996, p. 56)

## Energy Conservation: Contraindications of Oral Motor Exercises and Speech Drills

Lack of empirical evidence to support oral motor exercises (OMEs) in motor speech remediation.

"…the existing research literature provides insufficient evidence to support or refute the use of nonspeech OMEs". (McCauley, R. et al., 2009, 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". (McCauley, R. et al., 2009, p. 356)

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## Contraindications of Oral Motor Exercises and Speech Drills Continued

Bandaid on a gunshot wound approach – "Because of the pathopysiology 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)
http://www.asha.org/public/speech/disorders/ALSChallenge.htm

Fatigues muscles using non-purposeful tasks.

Unclear if exercise to fatigue may actually hasten neurologic deterioration.

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## Is There a Role for Exercise?

**(Plowman, E. 2015)**

- "…insufficient data to conclude that overuse weakness or functional decline actually occurs following exercise in PALS". (p.1155)
- 18 exercise-based intervention studies reviewed (1960-2014)
  - "Limb and respiratory exercise, applied early and at mild to moderate intensities may have a positive impact for maintaining motor neuron integrity". (p.1161)
  - "…recent data suggest that we need to consider the possible complementary role of mild forms of exercise…". (p.1161)

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## Speech Staging System for ALS

Stages:

1: No detectable speech disorder.

2: Obvious speech disorder with intelligible speech.

3: Reduction in speech intelligibility.

4: Natural speech supplemented with AAC

5: No useful natural speech.

(Yorkston, Beukelman, Strand & Bell (1999)

Additional Resource:

http://aac-rerc.psu.edu/index.php/files/list/type/1

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## 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.

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## Voice Banking versus Message Banking: What's the Difference?

- Voice Banking: creation of a synthesized version of human voice from a large number of recorded messages.
- Message Banking: digitally recorded words, phrases and/or sentences using natural voice, intonation and inflection.
  - Message Banking by Proxy
  - Legacy Messages/Story Banking

(Costello, J., 2014, Boston Children's Hospital Message Banking Examples from People with ALS)

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## Voice Banking/Message Banking Options

Voice Banking:

- Model Talker
- Acapela
- VocaliD

Message Banking:

- www.messsagebanking.com
- Creation of wav files

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## 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.

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## 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.
- Equipment options:
  - Voice amplifier
  - Palatal lift

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## 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).

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## Supplemented Speech Interventions Continued

- **Topic Supplementation** – speaker indicates the topic of the message.
- 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.

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## Supplemented Speech Interventions Continued

- **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.
- Limitations –
- Idiosyncratic.
- Not all messages are "gesturable".
- Impairments in upper extremity functioning are a limiting factor.

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## Supplemented Speech Interventions Summary

- Overall, dysarthric speakers' speech comprehensibility 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)

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## 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.
- Equipment options:
  - Voice amplifier
  - Palatal lift

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# 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)

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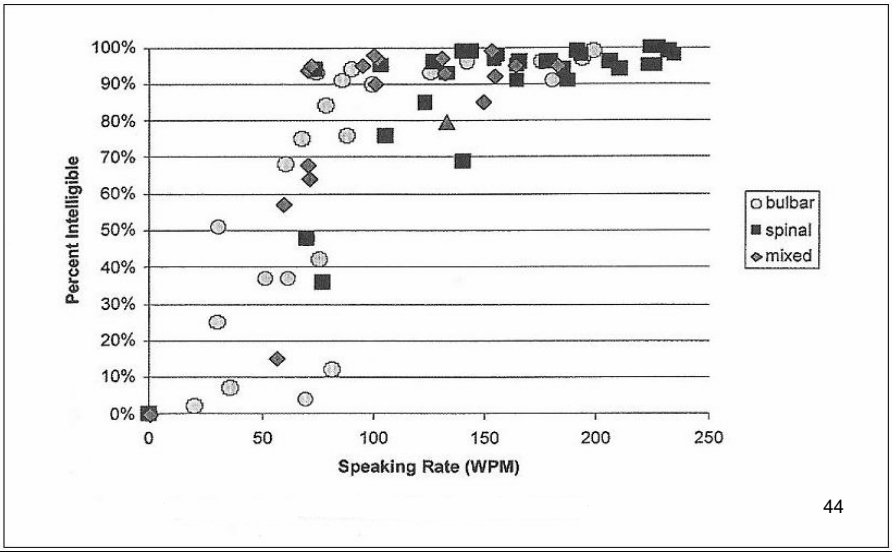
# Relationship Between Intelligibility & Speaking Rate

**(Ball, L. et al., 2002, p.234)**

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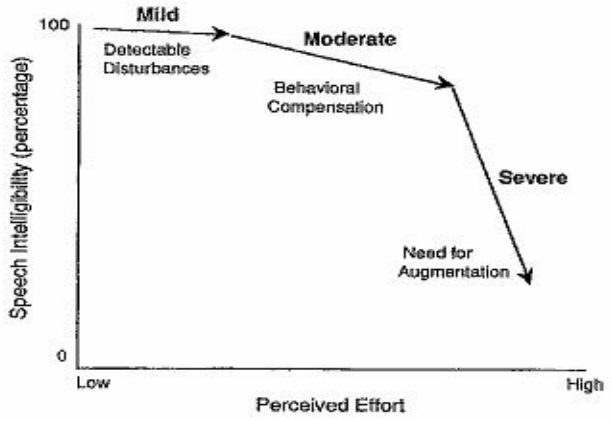
# Perceived Effort and Speech Intelligibility

**(Wilkinson, C. et al., 1995, p.142)**

**FIGURE 3. Hypothetical representation of changes in intelligibility and perceived effort as dysarthria associated with ALS progresses.**

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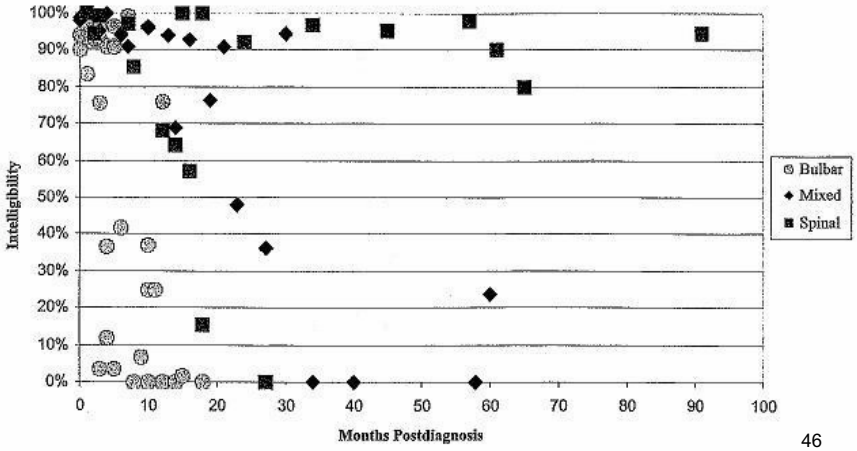
# Speech Deterioration: Intelligibility & # of Months Post-Diagnosis

**(Ball, L. et al., 2002, p.233)**

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## Timeliness of AAC Evaluations - "Reality Check"

- Insurance denial issues.
- 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)

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## 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.
- Equipment options:
  - Voice amplifier
  - Palatal lift

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# Voice Amplification

- **Benefits** –
  - Easy to use: no significant training needed
  - Inexpensive: prices range $50-$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.

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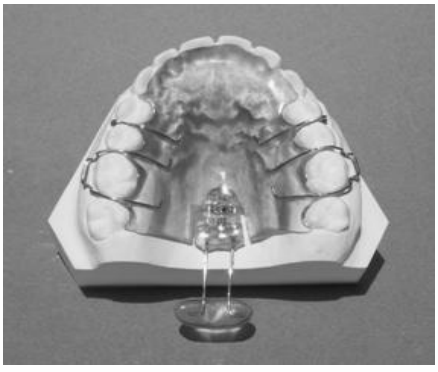
## Palatal Lifts



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## Palatal Lift Efficacy

Esposito, Mitsumoto & 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.

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## 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".
- Community prosthedontist support – ? variable or limited

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## Who might be a good candidate for a palatal lift?

Yorkston, Miller and Strand (2004) use the following guidelines:

- "Poor velopharyngeal function in the presence of relatively preserved lip and tongue movement
- Preserved ability to swallow saliva
- Adequate dentition to support the prosthesis
- 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)

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## 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.

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## 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.

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## Useful Websites

**ALS Information**

ALS Association: http://www.alsa.org/

MDA Association – ALS Division: https://www.mda.org/disease/amyotrophic-lateral-sclerosis

Northeast ALS Consortium (NEALS): http://www.alsconsortium.org/

Project ALS: http://www.projectals.org/

ALS Untangled: http://www.alsuntangled.com/index.html

**Patient/Caregiver Resources and Online Support Groups**

ALS Forums: http://www.alsforums.com

Patients Like Me - ALS/MND: http://www.patientslikeme.com/als/community

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# 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 consider contacting your local Neuromuscular Clinic (ALSA or MDA).

THANK YOU!!

Kim Winter: 860-827-1958 ext. 2035 kwinter@hfsc.org
www.hfsc.org

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