ALS and Augmentative Communication: Seeking Improved Outcomes through Early Engagement in Assessment, System Design and Implementation

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Dear John, Amanda, and Peggy,

I want to thank you for everything you did for my mother. Although she passed before she was able to share final days of the memory banking, I think our visits with you were an important part of her journey. As you know, it is a disease of constant loss, and all of us can only focus on that loss until our first meeting at Children’s with John. At that time we were able to discuss the technological Ramos available to my mom, but what we were really talking about was hope. An ability to be provided with motivation, passion, and the opportunity to hold onto a piece of who she was. A precious gift of the risk of empowering John, after that week I spent with you and the support I had with you was meaningful.

Sincerely,
[Signature]
**Program Mission:**
The mission of the ALS Augmentative Communication Program is to provide comprehensive augmentative communication/assistive technology assessment, trials and training to people with ALS from the time of diagnosis through the lifespan.

**Program Goal:**
"Our goal is to support communication and daily functional needs, sustain personal control and dignity, facilitate continued social and vocational goals and maintain quality of life through thoughtful implementation of solutions ranging from high technology to quick access/low tech tools and strategies. This is best accomplished by ACP-ALS clinicians constantly communicating and collaborating on how best to support patient-centered functional outcomes in the presence of changing physical abilities while providing support to a person with ALS and his/her family."

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Our team hopes to meet people as early as possible after diagnosis but remains eager to support people with ALS at *any time* during their journey.
### Types of ALS/MND

- **Sporadic** - the most common form of ALS in the United States - 90 to 95% of all cases.
- **Familial** - occurring more than once in a family lineage (genetic dominant inheritance) accounts for a very small number of cases in the United States - 5 to 10% of all cases.


### Onset

- **Bulbar**
- **Spinal**
- **Atypical**
  - Example: Brachial amyotrophic diplegia (man in the barrel): severe muscle involvement was confined to the upper limbs, predominantly the proximal portion and shoulder girdle, sparing the face and the legs until late in the disease's course or until the terminal stage.

### Bulbar onset

**What is it?**
- Bulbar ALS destroys motor neurons in the corticobulbar area of the brainstem in the early stages of ALS.
  - The corticobulbar area controls muscles of the face, head and neck.
  - Bulbar ALS usually progresses faster than limb onset

**How Common is Bulbar ALS?**
- observed in 20-30 percent of people with ALS.
  - Almost all people with ALS display bulbar symptoms at later stages

**Symptoms Affecting Speech**
- Changes in voice and speech: - Harsh, hoarse or strained voice.
  - Breathy speech pattern.
  - Poor articulation.
  - Decrease in range of pitch and loudness of voice.

**Other Symptoms**
- Spasms in muscles of the jaw, face, voice box, throat and tongue.
  - Inappropriate excessive laughing and crying.
  - Brisk jaw jerks.
  - Involuntary twitching in the muscles of the tongue.
  - Vocal cord spasms causing the sensation that air cannot be moved in or out.
Spinal onset

- initial symptoms may affect only one leg or arm. Individuals may have awkwardness and stumbling when walking or running. They may have difficulty lifting objects or performing tasks that require manual dexterity (e.g., buttoning a shirt, tying a shoe, turning a key). Eventually, the individual will not be able to stand or walk, get in and out of bed without help, or use hands and arms to perform activities of daily living, such as washing and dressing.
- 70-80% of patients, symptoms begin with limb involvement
- Eventually develop bulbar symptoms

Resource: Orphanet Journal of Rare Diseases 20094:3 DOI: 10.1186/1750-1172-4-3

Spinal onset (cont’d)

- Upper motor neuron involvement include spasticity and exaggerated reflexes
- Patients with upper limb onset have twice the likelihood for onset in the dominant arm, compared with the nondominant arm
- Symptoms of lower motor neuron degeneration include muscle weakness and atrophy, muscle cramps, and fasciculations

What to expect when coming to our ALS Augmentative Communication Program
As appropriate, Speech-Language Pathology will:

- introduce strategies to minimize fatigue associated with speech including: strategies to enhance intelligibility or preserve energy, and may introduce varied voice amplifiers.
- May introduce our model of Message Banking and/or options for Voice Banking,
- partner with patient and family to create – over time – custom quick access communication tools
- Introduce and assess various communication technologies to support face to face communication as well as communication through internet/telephone.
- Establish and coordinate evidence based trials
- assess and provide call systems to meet individual needs.
- Provide partner training

Based on assessment of current voluntary motor abilities, Occupational Therapist/AT specialist:

- identify adaptations and tools to facilitate continued physical access to daily activities. A wide spectrum of options exist, ranging from minor modifications to one’s computer keyboard and mouse to hands free control of a computer, tablet and smartphone.
- In addition to supporting hand function as much as possible, voluntary movements of one’s eyes, head and feet are explored to minimize the overuse of any one muscle group.
- Accommodations to minimize fatigue and facilitate function often include a combination of:
  - positioning mounting adaptations,
  - low and high tech adaptive pointers,
  - alternative computer mice and switches
- In addition to facilitating one’s access to written and spoken communication, email, the Internet and social media, options for independent access to reading, television operation and other leisure time activities can also be addressed.

Also provide:

- Home-based services may be available when patient can no longer travel to the center.
- Tele-support
- Web based training modules (late 2016)
- Web based downloadable templates (late 2016)
Begin with THANK YOU
to many extraordinary people with ALS
Opening statement:

“My goal is to waste your time”

AAC/Speech Pathology Protocol of Assessment Considerations

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Boston Children’s Hospital ALS Aug. Com. Program
**Speech strategies**

- Pacing/segmenting with breath control
- Breathing awareness (diaphragmatic vs. clavicular)
- Reduce gravel with quieter voice (in concert with amplifier)
- Over articulation (without strain)
- Economizing
- Stretching – NOT oral motor exercise/repetitive motion. Discuss issues of muscle recovery.
- Letter cueing
- Topic cueing
- Counsel on positioning/support
- Counsel on speech fatigue/over-use and difficulty with recovery

**Partner training**

- Identify communication partners/supports
- Share anecdotal feedback from people with ALS and families
- Share handout on “Guidelines to Communication Partners”
- Discuss strengths and major challenges with asking yes/no questions
- Discuss the pros and cons of prediction and permissions that should be in place.

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Boston Children’s Hospital ALS Aug. Com. Program
Amplification considerations
• Counsel regarding impact of speech efforts on fatigue
• Discuss pro-active approach (as appropriate) to preserving energy
• Introduce amplification options
• Identify microphone headset placement considerations with head movement

Often will be told:
“I can talk loud enough, I just get worn out by 2 in the afternoon and am too fatigued”

Speech production requires:
• Articulation
• Phonation
• Resonation
• Respiration
When I use the amplifier

Patient photo or video

Amplification while using bi-pap

Assessment of transdermal microphone options
Patient photo or video

Call system(s)/switch control

Home alone and calling 911 when speech is difficult

The Silent Call Procedure
If you need to call 9-1-1 and you are unable to speak for any reason, here is what to do:
Press if the 9-1-1 dispatcher asks you to call.
Press if you need fire.
Press if you need medical.

Source: MA State 911 Department and the Executive Office of Public Safety and Security
Mass.Gov
Phone app for emergency requiring no speech

Message Banking
- Introduce concept/definitions and idea of 'technology agnostic'.
- Practice recording with a hand held recorder to support high quality recordings 'in the moment'.
- Share clinical stories and outcomes and provide concrete examples.
- Provide full handout with definitions and thousands of examples from people with ALS.
- Download, playback, label and store audio files, providing guidance for improving quality if needed.
- Review potential technologies that could accommodate message banking across varied platforms.
- Provide person with ALS with their own recorder to take home and use to functionally record.

Message Banking .wav technology given to people with ALS
• set at 16/44 baud rate
• Must use wind guard
• Hold close to mouth for best quality
• Practice timing of push – speak – push

Message Banking with your own voice digitally record and store words, phrases, sentences, personally meaningful sounds and/or stories using your natural voice, inflection and intonation.

These messages are cataloged as wav files and may then be linked to messages in a variety of augmentative communication technologies or sound storage files. This will allow you to ‘retrieve’ a message and speak it in your own voice but does not allow you to create novel messages by spelling. If you have recorded individual words, you may combine those words to create unique messages, although the output will sound more staccato than your natural speaking.

TERMINOLOGY:

Legacy Messages are those messages, often delivered with unique intonation and prosody that are unique or particular to you. It may be a ‘trademark’ message you say or it may be a trademark delivery of a message that many people say. A legacy message does not need to be meaningful to the general population; instead it may have unique and personal meaning to only you and a loved one. Further, a legacy message does not need to be real words to be meaningful. It may be the way you clear your throat in a sarcastic manner to communicate “I told you so!” or it might be the invented pet name you have for a loved one delivered with your unique voice, intonation and prosody. Similarly, legacy message may be that stereotypical thing you say after your favorite sports team scores or it may be a unique greeting you deliver to friends. Those close to you may be helpful with identifying these Legacy Messages because sometimes they are so naturally part of socially relating with others, you may not even be aware you are “known” for them.
Currently: 64 page Message bank handout

64 page handout will be on new ALS website but can be found now at:

- http://www.childrenshospital.org/~media/messagebankdefinitionsandvocab201613.ashx?la=en

Patient photo or video
Nancy

Bob – first time with ‘message bank’ messages

Patient photo or video

Patient photo or video

Patient photo or video
Until recently...
Edit one audio file into multiples

Audio conversion beta.1 (will be free to all)
Voice Banking

• Provide definition and description of process
• Provide examples of voices created

Voice Banking is a process of recording a large inventory of your speech that is then used to create a synthetic voice that approximates your natural voice.

Done successfully, this would allow one to spell and create unique messages and then speak them through a synthesizer that approximates one’s natural speech. The science behind this process continues to be in development with behaviors of available software. The ModelTalker is one such project from the University of Delaware Speech Research Lab. The website is: www.asel.udel.edu/speech/ModelTalker.html

◆ Model Talker
◆ Cereproc (Edinburgh Scotland)
◆ OKI Electronic Industry Co Japan
◆ Edinburgh Voice Banking and Reconstruction project
◆ Acapella project
◆ VOCALiD
The ModelTalker System was developed by the Nemours Speech Research Laboratory located at the Alfred I. duPont Hospital for Children with funding from the National Institute for Disability and Rehabilitation Research, the National Institutes of Health, and Nemours Biomedical Research.

Quick Access Encoding

- Standard Etran two-step encoding
- eye gaze and partner assist combination (AEIOU)
- Alpha – color encoding
- EyeSpeak board
Introducing partner assisted spelling.

Patient photo or video.

Partner assist scan.
Review of ‘body board’

Quick Access (non-electronic)

- Personal tabbed flip chart
- AlphaCore displays or others with direct selection by:
  - Hand
  - Stylus
  - Safe laser
On our new ALS webpage
Safe laser and core vocabulary

Wearable eye speak technology

http://lowtechsolutions.org

Amy Roman and Margaret Cotts

Boston Children's Hospital ALS Aug. Com. Program
Electronic encoding

- Minimize executive functioning demands for communicator and partner
- Provide a visual script/reminder of message progress

Writing strategies

- Notepad
- Notebook
- Boogie board
- Ipad/android – note apps
  - Finger
  - Rubber tipped stylus
  - Jot stylus
  - Apple pen
Speech Generating Device Assessment and trial(s)

**Language Features:**
- core vocabulary • phrase
- single words • Alphabet
- message organization (grid, list, taxonomic, contextual, etc.)

**Encoding strategies**
- Abbreviation expansion
- prediction (word, grammar, morphology) • letter stream prediction (Dasher)

**Access features (in concert with OT):**
- Direct selection (unaided)
- Direct selection (aided)
  - headmouse
  - eye tracking
- dwell, switch, blink
- Scanning
  - Single switch
  - Two switch
  - Use of switch interface for technologies
  - Software vs. tech access options within tech (accessibility features)

**Integration features:**
- Internet
- Telephone
- television
- text
- custom software
- system mirroring (Splashtop, Team Viewer, etc.)

**Other:**
- Language
- Text
- Symbols
- Synthesizer (and integration with environment such as ‘Alexa’)

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Boston Children’s Hospital ALS Aug. Com. Program
Predictable (Therapy Box):

Import audio in Predictable

NOTE: You are not able to hear the recorded audio until the end of the process

1. open app
2. click on the device
3. select the “right” tab
4. Move to the tab where you see the “Edit sharing” section
5. Click on the “edit sharing” button
6. Click on the “edit sharing” button
7. End and select the audio you would like to import into Predictable
8. Save the selected group of recordings
9. Format can be 40 or others
10. From the app go to phone
11. Select the phone you wish to play
12. Click on phone to stop
13. Send through email
14.敢于
15. The recordings will be saved in the email
16. End and leave the app

Ion Clipster
blue tooth speaker
## Wearable amplifier

![Image of wearable amplifier](image)

### Occupational therapy/Assistive Technology Protocol of Assessment Considerations

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**Physical Access Control Site assessment**

- Direct selection
- Non-direct selection
- Best control site (s) *don’t over fatigue one control site:*
  - Head, eyes, mouth, tongue, respiratory (sip/puff), voice, chin, shoulder, trunk, arm/hand, leg, knee, foot.
  - Pressure, excursion, range
- Neural access (neural switch), BCI

**General considerations for access selection:**

(1) the range and control of movement
(2) the amount of training and practice required to use and
(3) the short and long-term costs/benefits of using access method

**Access to iPad/Android tablets**

- Position of device
- Use of finger and/or stylus
- Voice typing
- Siri
- Mounting options
Phone Access

- Landline options
- Speaker phone options
- Smartphone use
- Hands-free cell phone use
- Switch scanning on iPhone
- Siri
- Mounting options

Call system/attention signal

- Commercial wireless doorbell
- Switch-adapted attendant alarm
- Baby monitor
- Portable speech output device with or without switch

Environmental control

- Enlarged TV remote controllers
- Switch access to TV functions, lights, fan
- Voice control for TV functions, lights, fan
- Control through SGD
Access to books (hardcopy or digital)

- Kindle/Nook/iBooks
- Hardcopy books/ book holders
- Page turners
- Audio books

Computer access: keyboard

- Built in accessibility features
- Keyboard/key size
- Ergonomic keyboards
- Forearm supports
- Typing aids
- Word prediction software
- Onscreen keyboard software

Computer Access: mouse

- Customizing computer mouse settings
- Adaptive/alternative cursor control options
- Hand –based
- Head-based
- Foot-based
- Eye-based
- Auto click software
- Switch click options
Computer Access: *speech*

- Speech recognition software
- Dictation strategies to improve software recognition
- Built in commands
- Custom commands
- Voice mouse controls

Speech Generating Device Access

- Touch screen
- Stylus and stylus holders
- Keyboard
- Different computer mice
- And/or trackball
- Mouse
- Headtracking access (head mouse, gyro mouse, etc.)
- Adapted mouse
- Switch scanning
- Eyetracking access
Brain Computer interface

- Projects with which we are currently affiliated:
- Oregon Health Science Project RSVP
- National Ctr for Adapted Neuro Technologies Wadsworth Ctr.

Mounting/positioning

Tele-treatment
Augmentative Communication Program at Boston Children's Hospital

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