



AAC Treatment for Persons with Primary Progressive Aphasia (PPA)

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Goals for afternoon:

Participants will:

1. Become familiar with language symptoms of PPA;
2. Formulate AAC goals for language intervention in adults with PPA;
3. Understand clinical research implications for intervention decisions related to adults with PPA.

- We are the “raft” after the “wreck.”
(LaPointe)
- AAC are the paddles for the raft. (MFO)

What is PPA?

1. A degenerative language disorder.
2. A language disorder that does not easily fit into the classical aphasia typology.
3. A syndrome, often followed by cognitive decline, that has been described with 3 variants.

Diagnostic Criteria for PPA

Mesulam, M. *Annals of Neurology*, 49 (4), April, 2001

1. Insidious onset and gradual loss of word finding, object-naming or word-comprehension skills in spontaneous conversation;
2. ADL limitations attributable to language impairment, for at least 2 yrs after onset;
3. Intact premorbid language skills;
4. Absence of significant apathy, disinhibition, forgetfulness for recent events, visuospatial impairment, visual recognition deficits or sensory-motor dysfunction within initial 2 yrs of L impairment;

5. Acalculia & ideomotor apraxia may be present in first 2 yrs.

6. Other domains possibly affected during 2 yrs, but language most impaired fn.

7. Absence of specific causes (i.e., stroke, tumor, infection, metabolic disorder) on neuroimaging.

Dx Characteristics

- Age of onset 40-75 years old, mean onset age of 60 years.
- Preponderance of male patients

PPA is a clinical syndrome which may overlap with

- Alzheimer's disease
- Frontotemporal dementia
- Corticobasal degeneration
- Dementia-lacking-distinctive-histology (DLDH)
- CJD
- ALS
- ACD (Asymmetric cortical degeneration; Caselli, 1995)
- Pick's disease

Kertesz & Munoz, Amer. J. of Alzheimer's Disease. (2002), 17(1).

3 variants of PPA

- Nonfluent progressive aphasia (NFPA)
 - *PPA with agrammatism*
- Semantic dementia (SD)
 - *Fluent progressive aphasia*
- Logopenic progressive aphasia (LPA)
 - *PPA with comprehension deficits*

NFPA: nonfluent progressive aphasia

(most common type of PPA in an AAC clinic)

- *Anomia* or “trouble thinking of or remembering specific words when talking or writing”.
- Initial “empty” speech with preserved melody and fluency but little information.
- Slow, hesitant and labored speech frequently punctuated by long pauses and filler words, early symptoms of agrammatism.
 - Simplification (generic words for specific concept)
 - Circumlocutions
 - Substitution by fillers (“thing,” “Whachamacallit”)
 - Phonemic paraphasias
- Marked increase in speech errors, early symptoms of a progressive apraxia of speech.
- Relatively preserved single-word comprehension with later difficulty comprehending complex syntactic structures.
- Stronger oral reading than generative language skills.

Progression of disease varies

- Yes/No confusion for responses
- Apraxia of Speech
- Agrammatism -> Mutism
- Written language generation often mimics spoken language generation.

SD (semantic dementia)

- Fluent, grammatical speech;
- Confrontation naming deficits (often word knowledge can be accessed through visuo-perceptual route);
- Surface dyslexia;
- Deficits in word comprehension (2-way naming problems); *“In time, even the most common words fail to be decoded and the comprehension of conversation becomes impossible, although visual recognition of objects and faces remains relatively preserved” (Mesulum, 2001).*
- Later connected speech includes neologisms and semantic paraphasias.

LPA: Logopenic progressive aphasia

- Word finding difficulties within fluent speech;
- Decreased output but relatively preserved syntax and phonology;
- Combination of the dysfluencies of NFPA with the semantic comprehension deficits of SD.

“The principal function of language is to label, categorize, and communicate thoughts through the mediation of arbitrary symbols (words).”

Damage to any part of the language network can interfere with word usage and word finding. Such anomia deficits provide sensitive markers for dysfunction within the language network. Consequently, anomia emerges as a nearly universal finding in the early stages of PPA.”

Mesulam, M. (2001). Primary progressive aphasia. *Annals of Neurology*; 49: 425-432.



Some dx drama.....

Within neurologic syndrome identification:

- PPA falls under the “Neary Criteria” for FTLD: Fronto-temporal lobar dementia (Neary, D, et al. *Neurology*, 1998).
- Kertesz et al consider it a variant of Pick’s disease (Kertesz, A, et al. *Neurology*, 2000).
- Confusion on term “semantic dementia.” 3 distinct variants (Gorno-Tempini, ML, et al. *Annals of Neurology*, 2004).

Concomitant symptoms reported in association with PPA: Speech and Language

Rogers & Alarcon (2000). AAC for Adults with Acquired Neurological Disorders.

- Agrammatism
- Alexia
- Anomia
- Apraxia of speech
- Dysarthria
- Dysgraphia
- Dysprosodia
- Echolalia & palilalia
- Hoarse voice quality
- Impaired aud. Comp.
- Impaired repetition
- Impaired spelling
- Impaired syntax
- Mutism
- Neologisms
- Phonemic paraphasias
- Pure word deafness
- Reduced sentence length
- Semantic paraphasias
- Slow speaking rate
- Staccato speech
- Stuttering
- Telegraphic Speech
- Verbal hesitancy & long phrases

Concomitant symptoms reported in association with PPA: Cognitive & Sensory

Rogers & Alarcon (2000). AAC for Adults with Acquired Neurological Disorders.

- Acalculia
- Amusia
- Dressing apraxia
- Hemianopia
- Hypethesia
- Impaired executive function
- Impaired nonverbal functions
- Impaired verbal memory
- Perseverative behavior
- Poor recall
- Prosopagnosia
- Short-term memory deficits
- Visual agnosia

Proposed Stages of Intervention during the Neurodegenerative Language Process: NFPA

- I. No noticeable interference with generative language but some word finding problems;
- II. Detectable language lapses with hesitations and dysfluencies;
- III. Reduction in language use leading to behavioral strategies and introduction of low tech AAC (circumlocutions; paraphasias; simplification; agrammatism)
- IV. Use of AAC tools and other techniques;
- V. No functional language.

PPA presents a clinical conundrum:

Language behavior is truly variable,
and we don't know if we are
looking at impairment of linguistic
competence or performance.

Communication Treatment Goals

- ***#1: To compensate for progression of language loss (NOT stimulate the language system to regain skills).***
- ***#2: To start early. Begin compensatory treatment as soon as possible. Be proactive so patient can learn to use communication strategies and tools.***
- ***#3: To include primary communication partners in all aspects of training, with outreach to multiple partners.***

The Treatment Challenges:

- 1. To put the patient's residual lexicon **visually in front of him** so that the patient can participate in daily activities as language skills decline.*
- 2. To engineer the environment to support successful communication.*



This is where AAC comes in....

You should be asking yourself:

- **Where, on the natural communication continuum, can we intervene first with AAC?**
- **treatment for the person with PPA and their partners?**
- **How do AAC strategies and devices change with progression of the syndrome?**

Enter REKNEW-AD

REKNEW-AD

- Reclaiming
- Expressive
- Knowledge
- in Elders
- With
- Alzheimer's disease



Premise for REKNEW-AD research

- Pairing external aids with familiar and spared skills (such as page turning, reading aloud, personal information) maximizes a person's opportunity for success.
- These skills are based on intact procedural and autobiographical memory.
- Symbolic representations may serve as semantic primes to stimulate lexical retrieval during conversation in moderate Alzheimer's disease (modAD).

- 3-year research project funded by NIH and NIDRR, with pilot funds from the Oregon Tax Check-off Funds.
- PIs: Melanie Fried-Oken and Charity Rowland
- Study 1 Question: Do AAC supports improve conversation by individuals with moderate Alzheimer's disease?

Method



1. Determine subject's preferred topic and vocabulary;
2. Develop communication board;
3. Conduct 9 10-minute videotaped conversations in homes with and without the AAC device (conversational condition randomly assigned/visit).

Study 1 subjects with moderate Alzheimer's disease (N=30)

Diagnosis of probable or possible AD by a board certified neurologist;
Vision and hearing within functional limits; English as primary language;
Exclude those with prior neurological diagnoses or communication disorders.

Gender	23 Females	7 Males
Age	Mean = 74 yr.	Range = 50-94
MMSE (0-30)	Mean = 12	Range = 5-18
CDR (0-2)	Mean = 1.73	Range = 1-2
FLCI (0-88)	Mean = 61	Range = 27-85

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**Study 2 Question: Do AAC supports
*combined with spaced retrieval
priming exercises* improve
conversation by individuals
with moderate Alzheimer's disease?**

*Added PPA as more patients
showed up in the OHSU AAC
clinic.*

Wayne

- 62 year old man;
- Retired HVAC technician;
- Completed high school;
- In-line flying airplane hobbyist (owned a hobby store for a while with his wife);
- Lives in urban Portland, Oregon with his wife.

Language screening

- BDAE: Complex ideational material (85%, errors on last 2 paragraphs); offered yes/no cards for response verification;
- BDAE: Writing to dictation: 5/10
- RCBA Functional Reading subtest: 8/10.

FLCI: 73/82 (between mild & moderate)

- Receptive score 39. Expressive score 34.
- Could not name half of pictures shown (hanger, harmonica, stethoscope, compass).
- Could not answer 2/3 open-ended questions (where would you like to go on a trip & what's your favorite holiday).
- Could not successfully write sentence about self (wrote nothing).
- Could not write all words to dictation (harmonica, compass, knocker, stethoscope)
- Could not pantomime 2 of 9 pictures shown (harmonica & hanger)

MMSE- 13/30

- Could not name year, season, date, day, month, or county of residence.
- Could not spell WORLD backwards.
- Could not recall 3 words from earlier in exam.
- Could not successfully repeat, "No ifs ands or buts."
- Could not write a sentence (free-writing about anything).

CDR (wife report)- 1 (mild)

Memory: only highly learned material retained; new material rapidly lost

Orientation: Severe difficulty with time relationships; Usually disoriented to time

Judgment & problem solving: Severely impaired in handling problems, similarities, and differences

Community affairs: unable to function independently at these activities although may still be engaged; appears normal to casual inspection

Home & hobbies: mild but definite impairment of function at home; more difficult chores abandoned; more complicated hobbies and interests abandoned

Personal care: needs prompting

SIB: 3 (out of 4)

He could not answer "what is this" when shown the picture of the spoon (although he could with the cup). He could successfully pantomime how to use both the spoon and the cup.

SALT variables to consider

N=12 control and 6 AAC conversations

- Total # utterances
- # non-productive utterances
- # explanatory utterances
- MLU
- # abandoned utterances
- # interrupted utterances
- Mean turn length in utterances
- Mean turn length in words
- Total # words
- Type/token ratio
- # on-target words
- # words first mentioned

Control vs. experimental 10-minute conversations: Utterance variables

SALT variable	CONTROL	AAC
Total # utterances	67.33	78.5
# non-productive utterances	31.25	37.83
# explanatory utterances	12.5	13.67
# abandoned utterances	6.0	7.5
# interrupted utterances	2.08	3.3
# 1 word utterances	21.75	22.67
Mean turn length in utterances	1.256	1.38
Mean turn length in words	6.245	6.76

Lexical variables

SALT variable	Control	AAC
Total # words	441	506
MLU in words	4.94	4.97
Type/token ratio	.3950	.3733
# on-target words	332	383
# words first mentioned	101.75	110
# different word roots	128.67	140.5

What do the numbers mean?

- With AAC support, we see more words and utterances.
- With AAC support, we see longer turns (in words and utterances).
- With AAC support, we see more non-productive, abandoned and interrupted utterances, but these may be permitted because of joint references.
- The AAC support provides a non-verbal symbolic joint reference that reduces TTR and increases 1 word responses.

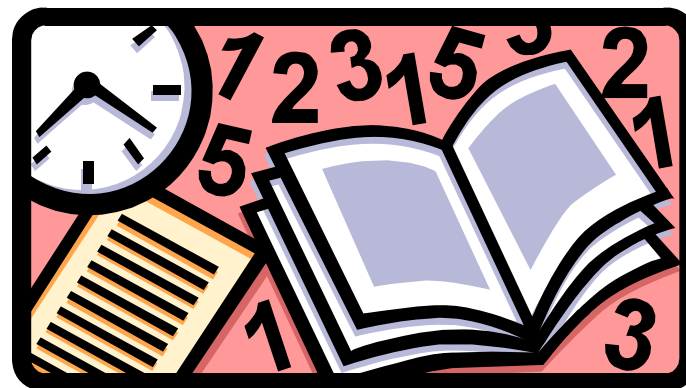
Tool use will vary according to the demands of the situation (*performance??*) and the user's abilities (*L competence??*)

- Consider language use variables:
 - Communication needs
 - Purpose of communication
 - Activity restrictions or opportunities
 - Level of communicator (independent, transitional, partner-dependent)

Visual representation for language

- Personal dictionary
- Calendars & Clocks
- Maps
- PDAs
- Family trees
- Pictures or photographs
- Drawings
- Remnant bags
- Augmented Input
- Continuum lines for conversation:

Hate _____ Adore



Low tech tools

- Customized communication boards
- Customized brag books
- Remnant bags/boxes
- Single message devices
- Talking photo albums

Personalized goals and tx

- “What would you like to be doing today that you are not doing? (Lasker)
- What goals would you like to pursue?
- How do you feel about your day? (Murphy,2000)
- How do you feel about your living situation (QofL) (Murphy and Gray, 2006)

Talking Mats: www.talkingmats.com



What is Talking Mats?

- A visual framework using picture symbols to help people with communication impairments to interact more effectively.
- A supported means to help individuals with choice making, goal setting, sharing opinions, and directing individual options.
- An interactive resource that uses 3 sets of picture symbols (topic, options, visual scale) with a textured mat.
- Presents topics in a structured, consistent and visual means for both comprehension and expression.

High tech tools

- Dynamic display devices with customized messages:
- Dynavox V or M3
- Words+ Say It
Sam Tablet SM1



- Digitized devices with hard copy pages (Saltillo Bluebird II or VocaFlex)



- Talking Photo Album (Augmentative Communication, Inc.)



Messaging

- What messages to include in tools?
- Svoboda: 100 autobiographical memories for elders
- Story telling: 87% of adult conversation is reminiscence and chatting

Partner training is an essential component of A

- To identify vocabulary for external lexicon.
- To support use of tools in familiar communication settings.
- To identify new opportunities for communication with tools.
- To offer or confirm choices.
- To initiate conversation during late stages of PPA.

Rogers, MA & Alarcon, NB. (1998). Dissolution of spoken language in primary progressive aphasia. *Aphasiology*.

Impairment 1993 -> 1997	Intervention Unassisted -> Assisted
<p>Increased frequency of speech errors</p> <p>Increased severity of apraxia of speech</p> <p>Difficulty accessing phonologic form</p> <p>Decline in appropriate syntax use</p> <p>Difficulty accessing semantic information</p> <p>Decreased access to orthography</p> <p>Auditory comprehension declines</p> <p>Reading declines</p> <p>Nonvocal</p>	<p>Develop long term Probes</p> <p>Pacing, syllable segmentation</p> <p>Decrease fillers, word retrieval</p> <p>Identify topics and key words</p> <p>Gestures, writing, drawing</p> <p>Involve communication partners</p> <p>Communication book</p> <p>AAC device</p> <p>Partner-focused guided communication</p>

www.brain.northwestern.edu/PPA*

- PPA Newsletters from 1996 (on line)
- Join mailing lists
- Connect to PPA databases
 - Clinician search and database
 - PPA literature database
- Question and answer archive
- PPA Family Support Group

- ***maintained by The Cognitive Neurology & Alzheimer's Disease Center at Northwestern University, Dr. M. Mesulam**

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AAC treatment in PPA: Workgroup Discussion

Tx dependent on level of communication ability

- Independent communicator
- Transitional communicator
- Partner-dependent communicator

Communication considerations

1. Behavioral strategies
2. Low tech techniques
3. High tech techniques
4. Partner-based strategies

Jack

- 70 years old
- Retired aeronautics engineer
- Built minutemen missiles and was consultant to U.S. military and private industry
- Moved to Portland, Oregon 1 year earlier, to be near daughter's family

Test scores

- **BDAE: Complex ideational** material-100% with yes/no cards
- **BDAE: Writing to dictation** – 8/10 with recognition of errors
- **RCBA Functional reading subtest:** 10/10
- **MMSE-** 29 (out of 30) Only recited 2/3 words remembered from a few questions earlier in the exam.
- **CDR-** 0 (no impairment)
- **FLCI--** 80/82; Expressive score 39. Receptive score 41. (Couldn't name stethoscope when shown a picture, did not correctly spell a few words to dictation (harmonica, knocker, stethoscope), and when asked to write a sentence about himself, he wrote, "I a minuteman millse guidance expert.")
- **SIB-** 4 (out of 4)

Recent references

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Murphy, J. (2000). Enabling people with aphasia to discuss quality of life. *British Journal of Therapy and Rehabilitation.* 7(11) 454-458.

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“Well, I could use this board to talk from breakfast to hell and back!”

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