PPA Treatment Update: Standardizing Care for Nonstandard Aphasia ASHA 2015

M.L. Henry¹, A. Mooney², D. Morhardt³
¹Department of Communication Sciences and Disorders, University of Texas, Austin; ²Oregon Health and Sciences University; ³Northwestern University

Abbreviated version_2016_OHSU only

Primary Progressive Aphasia (PPA)

- Slowly progressive aphasia caused by neurodegenerative disease
 - There must be no focal lesion (e.g., stroke)
- Most prominent clinical feature is difficulty with speech/language
- These deficits are the principal cause of impaired activities of daily living
- Often affects individuals <65 years

(Mesulam, 2008; Gorno-Tempini et al., 2011)

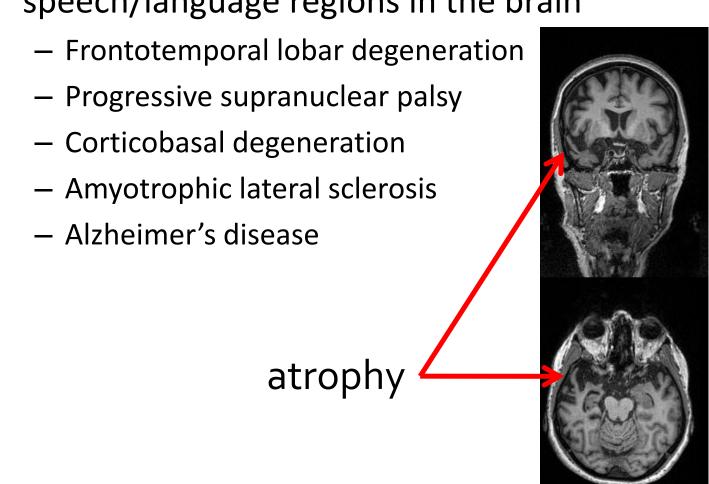
Exclusion Criteria for PPA

- Pattern of deficits better accounted for by other non-degenerative nervous system or medical disorders
- Cognitive disturbance is better accounted for by a psychiatric diagnosis
- Prominent initial episodic memory, visual memory, or visuo-perceptual impairments
- Prominent initial behavioral disturbance

Modified from Mesulam, 2003

PPA Etiology

 Caused by neurodegenerative disease that affects speech/language regions in the brain

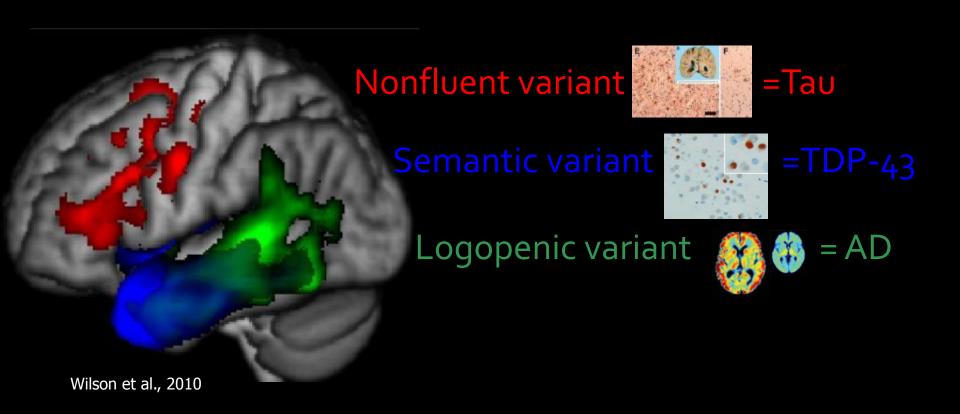


Deficits tend to arise in patterns: 3 variants of PPA (Gorno-Tempini et al., 2011)

- Nonfluent variant
 - Impaired syntax and/or motor speech (AOS, dysarthria)
- Semantic variant
 - Impaired semantic processing
- Logopenic variant
 - Impaired phonological processing (phonological loop)

FTD

These variants linked to underlying patterns of atrophy caused by different diseases



	Speech-language core characteristics	Speech-language associated characteristics	Typical imaging findings	Predicted neuropathology
Nonfluent PPA	One of the following must be present: 1) agrammatic production 2) effortful, halting speech with speech sound errors, including distortions, deletions, insertions, substitutions, transpositions (consistent with apraxia of speech)	Two of the following must be present: 1) agrammatic comprehension 2) spared single-word comprehension 3) spared object knowledge	left fronto-insular atrophy, hypoperfusion, or hypometabolism	Таυ
Semantic PPA	Both of the following must be present: 1) impaired confrontation naming 2) impaired single word comprehension	Three of the following must be present: 1) poor object knowledge 2) surface dyslexia/dysgraphia 3) spared repetition 4) spared grammar and motor speech	anterior temporal lobe atrophy, hypoperfusion, or hypometabolism	TDP-43
Logopenic PPA	Both of the following must be present: 1) impaired naming in spontaneous speech or to pictures 2) poor repetition of sentences and phrases	Three of the following must be present: 1) phonological errors in speech 2) spared single-word comprehension 3) lack of agrammatism	left posterior perisylvian/ temporo-parietal atrophy, hypoperfusion, or hypometabolism	Alzheimer's disease

Gorno-Tempini et al., 2011

Nonfluent PPA: Core Clinical Features (Gorno-Tempini et al., 2011)

One of the following must be present:

- 1) agrammatic production
- 2) effortful, halting speech with speech sound errors, including distortions, deletions, insertions, substitutions, transpositions (consistent with apraxia of speech)

Nonfluent PPA: Associated Clinical Features

Two of the following must be present:

- 1) impaired syntactic comprehension
- 2) spared single-word comprehension
- 3) spared object knowledge

Semantic PPA: Core Clinical Features

Both of the following must be present:

- 1) impaired confrontation naming
- 2) impaired single word comprehension

Semantic PPA: Associated Clinical Features

Three of the following must be present:

- 1) poor object knowledge
- 2) surface dyslexia/dysgraphia
- 3) spared repetition
- 4) spared grammar and motor speech

Third variant of PPA- not FTD

- Logopenic variant
 - Usually AD pathology

Logopenic PPA: Core Clinical Features

Both of the following must be present:

- 1) impaired word retrieval in spontaneous speech and confrontation naming
- 2) impaired repetition of sentences and phrases

Logopenic PPA: Associated Clinical Features

Three of the following must be present:

- 1) phonological errors in speech
- 2) spared single-word comprehension and object knowledge
- 3) spared motor speech
- 4) absence of agrammatism

PPA classification

+ naming impairment =SV, LV, NFV +single word comprehension impairment -single word comprehension impairment =LV, NFV

> +agrammatism and/or AOS -agrammatism and/or AOS =NFV=LV

SV= semantic variant of PPA LV=logopenic variant of PPA NFV=nonfluent variant of PPA

=SV

+ phrase/sentence repetition impairment =LV

Part 2

TREATMENT APPROACHES IN PPA

Understand the progressive nature of speech-language impairments

- Unlike stroke, in PPA, speech-language abilities gradually decline
- Initially, communication difficulties are the only cause of limitation to ADLs
- Ultimately, concomitant cognitive and motor difficulties develop

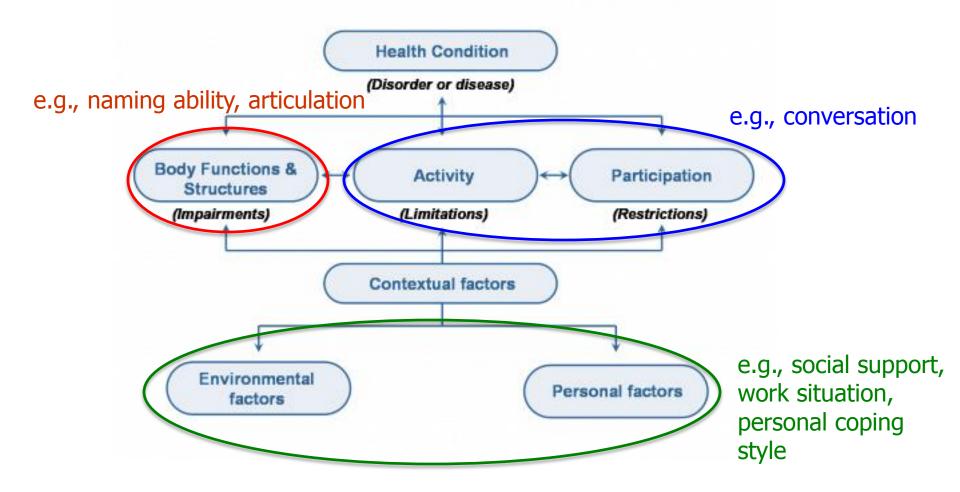
Over-arching goal: Functional communication

- Maximize communication at each stage of illness
- Consider the individuals in the context of their environment
- Tailor treatment approach with current status;
 take into account likely progression

Ideal model: Staged treatment approach

- Goals evolve with symptom progression
- 3 stages/phases
 - I. Restitutive
 - II. Shift toward aided approaches
 - III. Environmental support and partner training
- Address all levels of ICF model
 - Body structure/function
 - Activity/Participation
 - Contextual factors

WHO's International Classification of Functioning, Disability, and Health



Non-linguistic considerations that may affect treatment outcomes

- Changes in cognition
 - Memory
 - Executive functions
 - Visuospatial processing
- Changes in behavior
 - Social and emotional changes
 - Disinhibition, apathy
- Motor changes
 - Weakness, incoordination
 - Limb apraxia

Concomitant cognitive impairments

Cognitive Domain	PPA Clinical Syndromes: Degree of Impairments			
	lvPPA (Logopenic Variant)	nfvPPA (Nonfluent Variant)	svPPA (Semantic Variant)	
Calculations Numerosity Arithmetic Facts Complex Calculations	xx xx	х	х	
Executive Functions Task Switching Echoic and Working Memory Cognitive Control and Inhibition Phonemic and Semantic Fluency	(X) XXX X	XX X X X	(X) X	
Language and Speech Syntax/Grammar Motor Speech Production Confrontation Naming Fluency Reading Repetition Single Word Comprehension	XX XX XX XXX	XX XX X XXXX (X)	xxx xx xxx	
Memory Consolidation Verbal/Auditory Memory Visual Memory	XX X	(X)	X X	
Visuospatial Functions Visual Localization Visuoconstruction Face Perception	X X		(X)	

Displays the neuropsychological impairments noted in each PPA syndrome, grouped by cognitive domain. Level of impairment is denoted by an 'X', with 'XXX' suggesting severe impairment and likely a cardinal cognitive feature, 'XX' implying mild to moderate impairment, and 'X' signifying subtle to mild impairment. Parenthetical indicators [e.g. (X)] suggest that subtle difficulties may be noted, or are inconsistently evident in these patients.

PHASE I: RESTITUTIVE APPROACHES

Restitutive treatment for speech and language impairments in PPA

Background

- Despite considerable recent progress understanding the neurobiology of PPA, little research effort directed at behavioral rehabilitation
- Pessimism on the part of clinical professionals and third party reimbursers
- Case reports and single-subject experimental research provide descriptions of behavioral intervention in <30 individuals with PPA

Treatment for speech and language impairments in PPA

- A number of treatment approaches appear promising (see Rising et al., 2014 for a review)
- Treatment effects can be substantial and lasting
- Can result in changes in speech-language behaviors and observable changes in neural processing (fMRI)

Restitutive PPA treatment by variant

- Vast majority of studies address lexical retrieval, but sentence production and written language also treated (for review, see Rising et al., 2014)
- Most research to-date with semantic variant
 - Modest, item-specific gains
 - Continued practice required for maintenance
- Few nonfluent/agrammatic cases in the literature
 - Several have shown significant improvement in naming trained lexical items (e.g., Jokel et al., 2009)
 - One study addressing agrammatism (Schneider et al, 1996)
 - One study addressing motor speech (Henry et al., 2013)
- Few logopenic (lv) cases in the treatment literature (Beeson et al., 2011; Henry et al., 2008b; Henry et al., 2013; Newhart et al., 2009)

PHASE II: COMPENSATORY/AIDED APPROACH

Treatment themes in PPA

- Timing: Start early & be proactive so person with PPA can learn to use communication strategies and tools as soon as possible
- Communication partners: training from the beginning and throughout
- Degeneration: Adjust treatment strategies over time and use multiple modalities to capitalize on patients' strengths

- Early intervention
- Individualized treatment
- Patient & Family Centered Care + Goals
- Partner Training
- AAC Approach
- Education/counseling

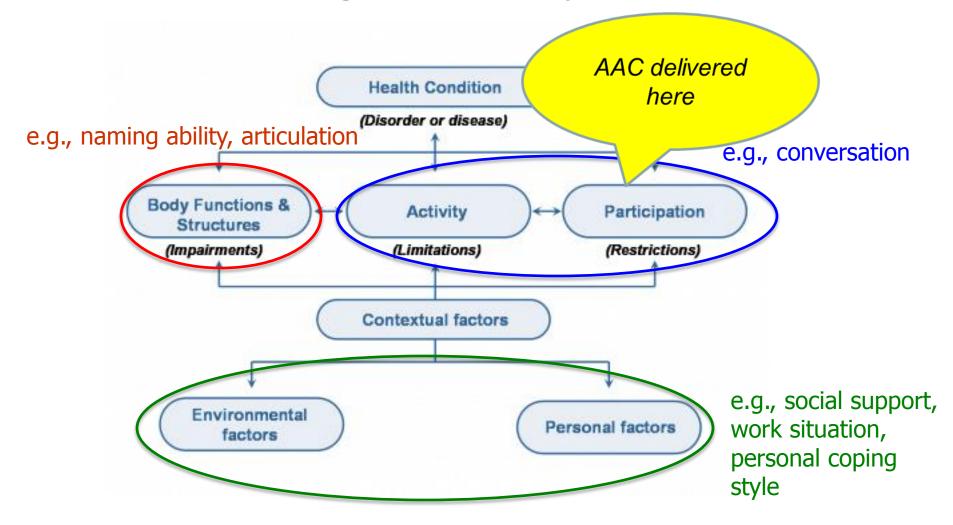
COMMUNICATION SUPPORT

"Anything that improves access to or participation in communication, events, or activities.

...includes strategies, materials, or resources that are used by people with impairments or by others who communication with people with impairments.

...It involves modifications in the environment around the person or to activities...also includes supportive attitudes that foster communication participation."

WHO's International Classification of Functioning, Disability, and Health



Restuitive

Improve speech intelligibility

Compensation

Compensate through use of tools, environmental and partner adaptation

Goal = return to previous levels of function

Goal = provide ways to remain engaged in daily activities with alternative approaches or tools

(Khayum, B., Wieneke, et al., 2012; Kortte, K. B., & Rogalski, E. J., 2013)

More on Assessment in PPA

- 1. language competence: articulation, fluency, syntax, grammar, word retrieval, repetition, comprehension, reading & writing
- 2. language performance: demands of environment & functional communication skills needed for different settings and situations

Social Networks Inventory

(Attainment Company)

Aphasia Needs Assessment

(Garrett & Beukelman, 2006) http://aac.unl.edu

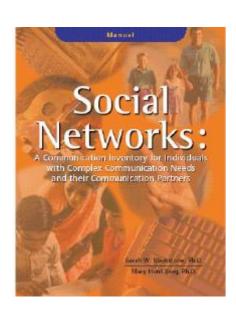
CETI: Communicative Effectiveness

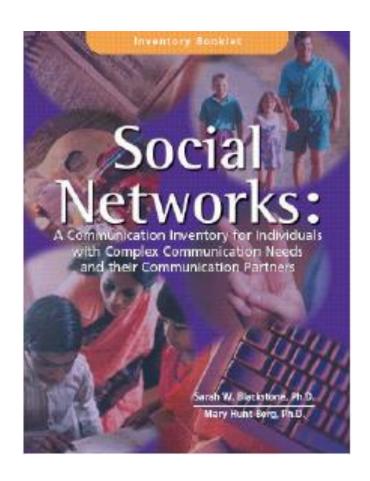
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(Lomas et al., 1989)

Social Networks:

A Communication Inventory for Individuals with Complex Communication Needs and their Communication Partners



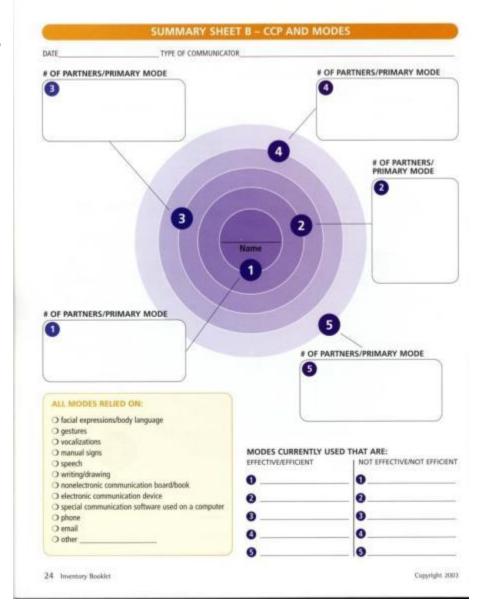


Circles of Communication Partners: Social Networks

(Blackstone & Berg, 2003)

Identify partners and modes:

- 1. Intimate partners (family)
- 2. Good friends
- 3. Acquaintances
- 4. Employees
- 5. Strangers



Considerations:

Partners

- Primary partner?
- Most skilled?
- Spends most time with ?
- Willing to learn new communication skills?
- Person most willing to teach others how to communicate with individual?

Modes

- Choice of modes is influenced by the situation, intent, content and individuals involved
- Performance is multi-modal
- Modes uniquely constrain types of information conveyed

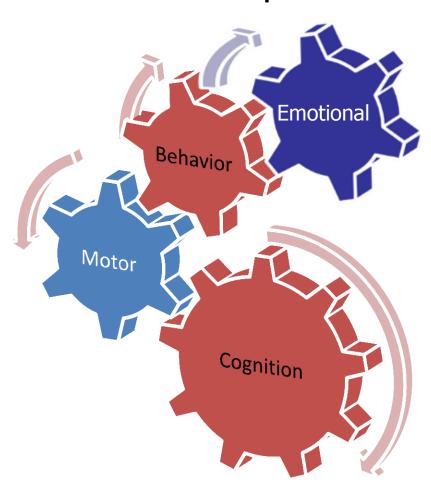
Ideal model: Staged treatment approach

- Goals evolve with symptom progression
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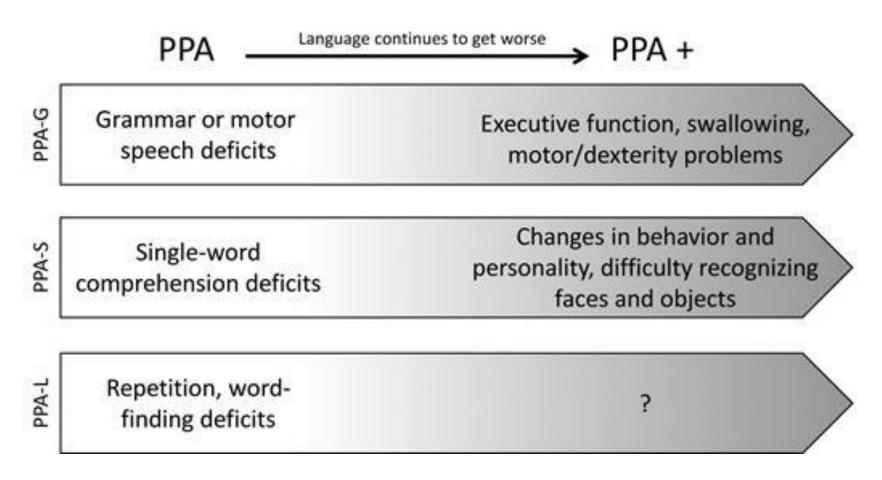
Staging PPA treatment: Fried-Oken, Rowland & Gibbons, 2010

Stage	Treatment	Partner Involvement
I: RESTORATIVE Detectable language lapses with hesitations, dysfluencies and word-finding difficulties	Education; behavioral strategies to support conversation. Introduction of low tech AAC.	Behavioral training: -how to ask questions -provide choices -alter verbal and physical environment to support communication
II: COMPENSATORY/AIDED Reduction in language use (circumlocutions, paraphasias, simplification, agrammatism)	Stage I + additional low tech AAC. Transition to other tools and techniques for multi-modal communication system, (mobile devices and SGD).	Device training: Partners learn message selection techniques and operations of each AAC tool.
III: ENVIRONMENTAL Little to no functional language	Environmental Support	Co-construction training: Partners lead successful interaction; support participation with multi- modal techniques.

Contributing factors to consider for treatment and compensation choice:



Progression of PPA to PPA+



Khayum, Becky, et al. "Thinking outside the stroke: Treating primary progressive aphasia (PPA)." *SIG 15 Perspectives on Gerontology* 17.2 (2012): 37-49.

Phase II: A shift toward aided approaches

- Expressive language is less efficient
- Verbal participation in all activities decreases
- Telephone use decreases or is avoided
- Conversations become imbalanced

Communication Supports



Unaided Approaches

(Natural modes and strategies)

- Speech
- Vocalization
- Gestures
- Eye gaze
- Body language
- Sign language
- Partner co-construction

Aided Approaches (Low tech and high tech tools)

- Paper and pencil
- Communication cards/boards and books
- Speech generating devices
- Mobile technologies
- Communication partner support
- Environmental modification

Consider communication demands

Settings

- Employment
- Home
- 1:1; Group
- Community event
- Partners
- Topics
 - Familiar vs. unfamiliar

Modes

- Telephone
- Face to face, spontaneous
- Written
- Electronic (text; email)



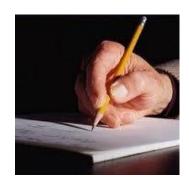




Low tech options

- Paper/pencil
- Communication books
- Communication passports
- Photo albums
- Pictures
- Newspapers
- Communication boards
- White board



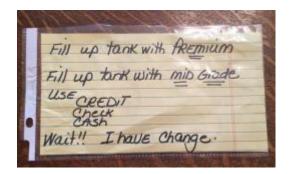






Low tech options

- Cards
- Remnants
- Written choice and continuum lines
- Paper and pencil
- Lanyards



Wong-Baker FACES Pain Rating Scale



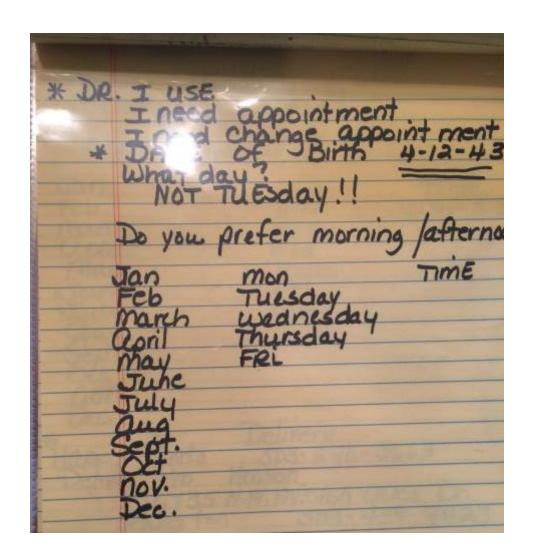
From Wong D.L., Hockenberry, Eaton M., Wilson D., Winkelstein M.L., Schwartz P.: Wong's Essentials of Pediatric Nursing, ed. 6, 5t. Louis, 2001, p. 1301. Copyrighted by Mosby, Inc. Registrated by Mosby, Inc.





Low tech options: scripts

- Can be prepared with conversation partner
- Used over telephone
- Order food, set up appt, ask directions
- Explain PPA, ask for support
- Personal information



High tech options

- Dedicated speech generating devices
- Mobile technology devices











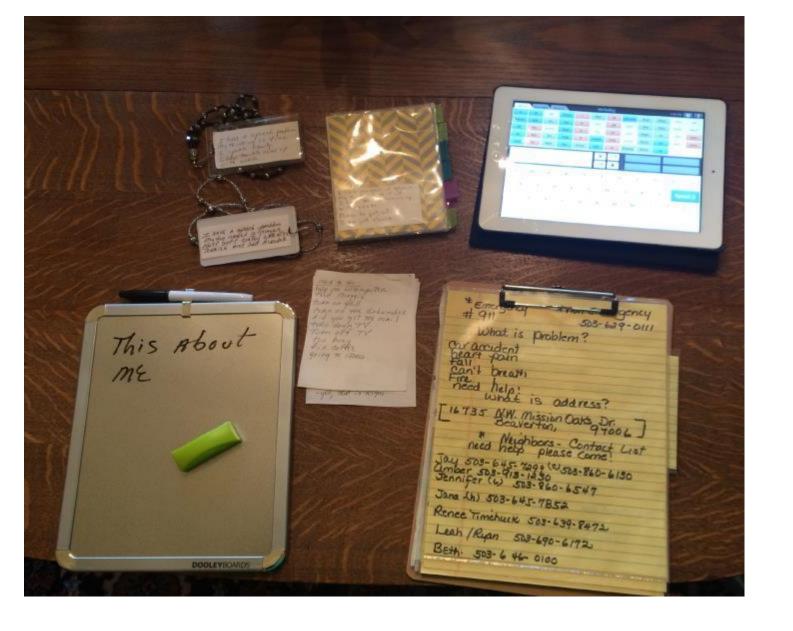
High tech options

Speech-Generating Devices

- Simple to complex
- Digitized or synthesized speech
- Individual can still use own speech with this supplementation
- As severity increases may be main mode (or not as cognition and fine motor decline)
- Many options: Lingraphica,
 Dynavox = \$8K; Go talk 20 = \$200

Mobile Technology

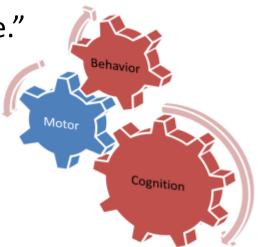
- Social acceptance
- Consumer empowerment in accessing assist
- Connections to social network
- Native applications
- Multiple applications
 - ProLoquo2Go
 - Lingraphica SmallTalk
 - Pictello
 - Scene and Heard



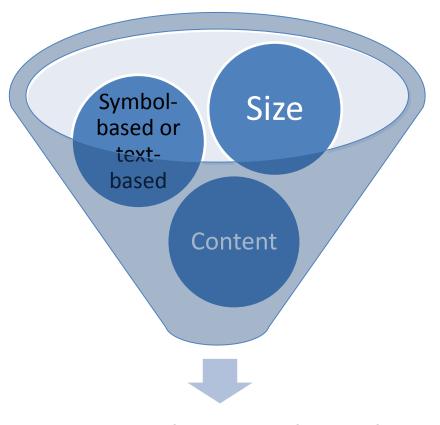
AH: an arsenal of options

User features to consider

- Previous experience with technology
- Support for training
- Partner's experience with technology
- Working memory abilities
- Vision and hearing abilities; fine motor skills
- Cognitive strategies and skills
 - "She can't seem to find the correct page."
- Motivation
 - "I bought this for mom to use."



Tool features to consider



Low Tech or High Tech

What AAC tools do participants report continuing to use?

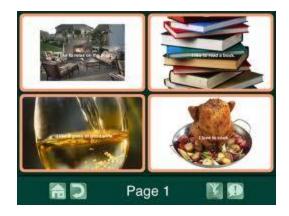
- Address book
- Ads
- Calendar
- Children's bible stories
- Comm. Board
- Comm. photo booklet
- Church bulletin
- Computer
- Cookbook
- Electronic photo frame
- E-mail
- Flashcards
- Gestures/ sign language/ pantomime
- Letters

- Magazine
- Mail
- Maps
- Museum brochures
- Newsletters (school, neighborhood)
- Newspaper
- Numbers
- Paper & pen/writing
- Photos & photo albums
- Post it notes
- Resident/staff directory
- Scrap book
- Show Me
- Skype

Compensatory/aided research background

Do AAC tools improve quality of conversations for individuals with degenerative language impairment?





A series of studies 2004-2015



- **Studies 1 & 2**: Do personalized low tech AAC boards in *controlled conversations with research assistants and with frequent partner (spouse, child, caregiver)* improve expressive communication?
- Study 3: Is there generalization and maintenance of AAC over 6 months?
- Study 4: Does use of mobile technology for language support improve conversation in people with PPA?

Low tech AAC study (Fried-Oken & Rowland, 2012)

Method

- 20 individuals with PPA
- 20 individuals with AD
- Personalized communication boards
- Trained in board use with RA and with spouse
- Scripted and natural conversations; 6 with and 6 without AAC

PPA Results

- Target words produced at initial prompt significantly greater with AAC
- Number of questions posed by RA or spouses to elicit target words significantly reduced with AAC

Interpretation of results



- Low tech AAC provides meaningful lexical support during structured conversations for people with PPA.
- Low tech AAC significantly reduces lexical scaffolding provided by the conversation partner.
- This approach should be part of a PPA treatment protocol.

Next Steps: mobile technology for PPA Sharing new information with/without AAC

Method

- Justification: "I can understand what he is saying when I start the conversation. But when Jim comes up to me and wants to tell me something, and I don't know the topic, I have no idea what he is talking about!"
- 6 adults with PPA (3 mild; 3 severe)
- Completed 3 activities with RA
- RAs populated GoTalk Now app with photos & speech
- Barrier task: Tell your spouse what you did this afternoon
- 3 conversations with no AAC support; 3 conversations with AAC

Pilot Results

- Significantly greater report of gist with AAC than with no support
- For subjects with severe PPA: Total words spoken with AAC increased significantly compared to speech only condition;

Current study in mobile technology for PPA:

Co-Construction of external lexica with mobile technology in PPA

This project combines AAC technology and innovative Natural Language Processing (NLP) techniques to develop and evaluate a novel intervention tool for individuals with language loss secondary to primary progressive aphasia (PPA).

We will develop a mobile technology app to support language co-construction and word finding skills in adults who are experiencing devastating and gradual language impairments.

OHSU PPA STUDY

Purpose: to find out if using an app on the iPad will help people with PPA to communicate more clearly.

Does having pictures and/or written words may help to tell a story?





Co-Chat®

Generates a lexical display, using NLP techniques, based on:

- photographs taken by user
- > comments from user's "social network"
- auto-curated list of related key words

Methods

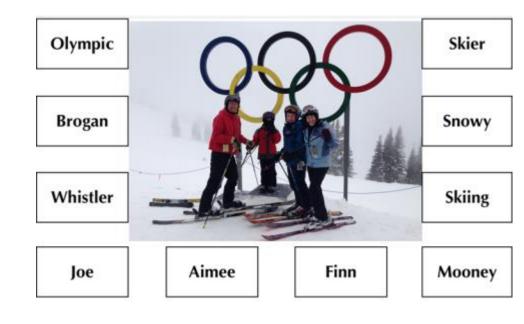
- Consent participant and communication partner in their primary residence;
- Assess language, cognition, vision/hearing
- 3. Train app use
- 4. Conduct an activity in the participant's home when conversation partner NOT present; photograph and send to "SN" for comment
- Comments analyzed by LM and generate lexical display with original photo

Participants engage in 6. meaningful conversations with familiar partners

Three conversation conditions

- ➤ No tech
- ➤ Photo only
- ➤ Co-Chat® Photo
 with words
 generated from
 Social
 Network/Language
 Model





How will we measure improvement?

Compared to no tech conversations:

Number and percent of target words used

PHASE III: ENVIRONMENTAL APPROACH

Natural environmental supports

- Pointing to weather pictures in newspaper to indicate time of day
- Using mail received from the bank to indicate questions about finances
- Flipping through pictures in photo book during a family visit
- Placing cue cards throughout environment
- Remnant boxes
- Native apps on mobile devices (weather, photos, calendar, maps, etc)

Clinical message: AAC WITHOUT TRAINING IS NOT AAC AT ALL!



Partner training is essential component of AAC for persons with PPA.

- > Role of partners paramount for success:
 - As person with PPA loses language, partner assumes more responsibility for interaction and message con-construction.
 - Evidence shows training is effective in improving communication activities and participation.
- To identify vocabulary for external lexicon.
- ➤ To support use of tools in familiar communication settings.
- To identify new opportunities for communication with tools.
- > To initiate conversation during late stages of PPA.
- > To help train other partners

Training Partners for Optimal Communication Strategies

Comprehension Facilitation

- Face to Face; get attention 1st
- Augmented input
 - with gestures
 - with pictures
- Speak slowly, allow to respond
- One direction at a time
- Written choice
- Yes/no choice
- Supplement Manage the environment
- To offer or confirm choices.

Expression Facilitation

- "Should I finish his sentences? Give him the words?"
- Support all forms of communication
- Count to 10 in your head before expecting a response
- Set up a way to "come back to that later."

www.reknewprojects.org -> Primary Progressive Aphasia -> Communication supports

Helpful Hints for Conversation

Use the examples below to help you think about how to begin a conversation, keep a conversation going, redirect the conversation, or to expand the conversation beyond one topic.

Request Details

- · Can you give me a specific example?
- How did that happen?
- Why did you go?
- · What were the names of the other people?

Request More Information to Expand the Conversation

- · Is there anything else you can think of?
- Tell me more about...
- · Had you done similar things?

Ask About Context

- Who else was there?
- What were you wearing?
- What color was it?
- · Who did you travel with?
- What did you eat?
- · How did the flowers smell?

Had you ever been there before?

Ask About Time/Sequence

- When did it happen?
- · What day of the week was it?
- Was it dark or light?
- · What time of year did it happen?
- · How long did it last?
- What happened next?

Ask About Place

- Where did it happen?
- · Were you inside or outside?
- What room were you in?
- Where were you sitting?
- · What sorts of things were around
- · Did you stay there or go somewhere else?

Acknowledge Any Response

- Yeah, I like it there too.
- You're right, she is a wonderful friend.
- · I remember doing that, and then
- · That was a long time ago, but what I'm really asking is...
- I'd love to talk more about that.



REKNEW Projects Child Development and Rehabilitation Center

Guidelines for Communicating with People who have **Communication Difficulties**

Remain Calm and Positive

- · Smile and remain interested even when conversation strays.
- . Keep a level head, a calm voice. remain as relaxed as possible.
- . Focus on what the person can do, not what they can't do.
- · Look for opportunities to support interaction.

Keep it Simple

- . Speak in short, concrete sentences.
- · Rephrase to keep topic focused when person is confused.
- · Respond immediately to communication attempts.
- · Provide clear choices between no more than two possibilities.

Be Polite

- . Make sure the person is willing to have a conversation.
- · Maintain eye contact (if culturally appropriate).
- · Reassure and support the person if stuck or frustrated.
- . Thank the person for having a conversation.

Support All Forms of Communication

- . Encourage and validate the use of any communication techniques.
- · Use pictures or other aids to help with word finding difficulties.
- · Encourage pointing and other gestures.
- · Encourage facial expressions.
- · Encourage writing and drawing.

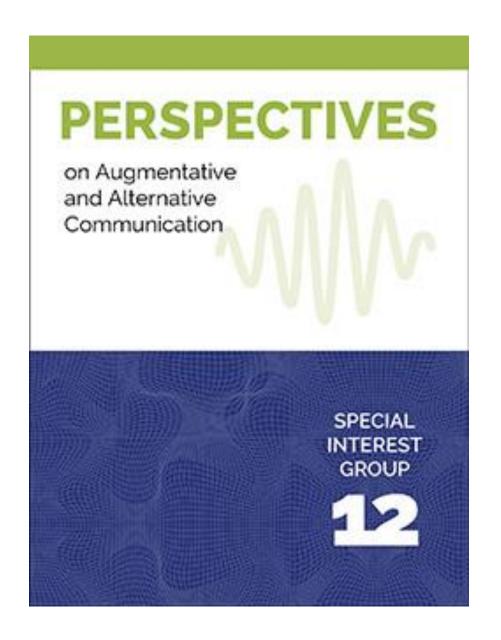
Reduce Frustration

- · Request more information on a topic if unclear.
- · Avoid quizzing just to get the "right" answer.
- . Do not directly contradict the person even if they are wrong.
- . Draw focus away from frustrating or embarrassing problems.

Be Aware and Informed

- · Monitor changing needs for communication support.
- Practice using all communication strategies yourself.
- · Role play with friends, family and therapists to understand how to handle communication breakdowns.

REKNEW © 2009



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Documenting successful outcomes for people with PPA

- Stability = progress
- Comprehension and production in any modality is acceptable
- PASS (Sapolski et al., 2008) 2011, 2014
 - Tool to rate symptom severity in a variety of domains (rather than signs of impairment)
 - 0-5 scale normal to severe from articulation to functional communication (similar to CDR)
 - Helps generate profile of strengths/weaknesses, disease progression and response to tx

Measuring progress:

- Given the degenerative nature of PPA, global
 measures of language function will not be sensitive to
 improvements in specific skills or behaviors that occur
 as a result of interventions
- Goals based on current assessment information; relevant to both caregivers & clients.
- Frequency counts
 - behaviors of interest
 - amount of assistance or cues required to perform a task
 - number and type of errors made during an activity
 - caregiver communication behaviors

RESOURCES

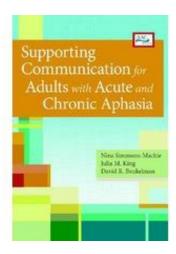
Webinar Series for PPA: http://www.brain.northwestern.edu/about/events/webinar.html

Northwestern, National Aphasia Association and the Association for Frontotemporal Degeneration developed a series of webinars for Speech and Language Pathologists (SLPs) who treat people with primary progressive aphasia (PPA).

- 1. Living with Primary Progressive Aphasia: Challenges Experienced by PPA Patients and Families and How SLPs Can Help!
- 2. Treatment for Persons with PPA: An Adaptable Communication Support Approach
- 3. The ABCs of PPA for SLPs: Clinical Attributes, Biology and Care of Primary Progressive Aphasia".

Book and website references for AAC

- www.aac-rerc.com: Research to practice AAC for PPA webinar (AAC Rehabilitation Engineering Research Center)
- http://memory.ucsf.edu
- www.brain.northwestern.edu (Cognitive Neurology and Alzheimer's Disease Center)
- Simmons-Mackie, King & Beukelman (2013) Brookes Publishing



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For more information:

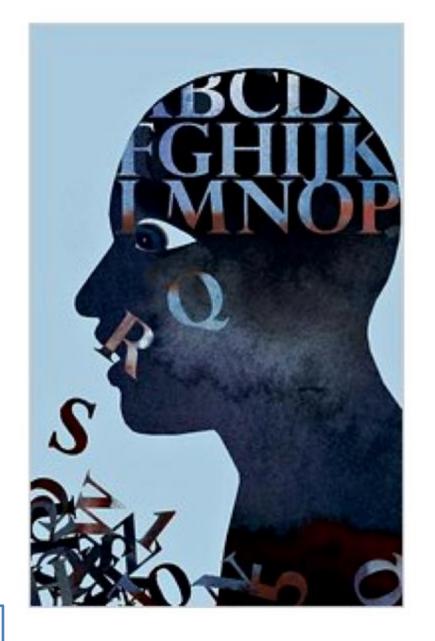
www.reknewprojects.org



A THIEF THAT ROBS THE BRAIN OF LANGUAGE

New York TimesMay 2, 2011 Jane Brody

http://www.nytimes.com/2011/05/03/health/03brody.html





www.aphasia.org



www.theaftd.org

IMPPACT

- IMPPACT International PPA Connection
 - http://www.ppaconnection.org
 - To foster international collaboration in PPA and serve as a compendium of patient-care resource related to PPA throughout the world.
 - Email <u>admin@ppaconnection.org</u> to access clinical videos related to PPA diagnosis.



