

A Life Participation Approach for Treatment for Individuals with Primary Progressive Aphasia

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What is Primary Progressive Aphasia (PPA)?

PPA is a clinical dementia syndrome caused by neurodegenerative brain disease. PPA is defined by progressive difficulties with language processing (i.e., aphasia) that initially emerge in the absence of other cognitive or behavioral changes.

Limitations of subtyping:

The PPA subtype is based on the prominent aphasia profile. **It is important to remember that many individuals do not neatly fit into one subtype, and may have symptoms from several different subtypes.** Also, PPA is a moving target. As the disease spreads throughout the brain, symptoms from the various subtypes will emerge. It is important to look at the person's clinical presentation of language strengths and weaknesses when developing a plan of care. The described subtypes are:

- PPA-L (logopenic variant) is characterized by the absence of grammatical difficulties, preserved single word comprehension and hesitant speech with pauses for word-finding;
- PPA-G (agrammatic/nonfluent) is characterized by agrammatism in speech, writing and/or comprehension, and preserved single word comprehension;
- PPA-S (semantic) by impaired single word comprehension and naming and preserved grammar.

A Person-centered Holistic Approach

The complexity of these diagnoses require a holistic approach, with the following variables taken into account when considering treatment strategies: (1) Clinical profile of language strengths and weaknesses, (2) Severity of symptoms, (3) individual's hobbies, interests and communication needs, (4) individual's motivation, along with emotional well-being, and (5) caregiver and family dynamics & involvement/support. A combination of home exercises, along with compensatory strategies may often be appropriate, with the recommended strategies frequently adjusted to meet a person's changing communication needs as the disease progresses.

Person-Centered Speech-Language Pathology Assessment: Flipping the Rehab Model

- What is the purpose of your evaluation?
 - Are you a part of a diagnostic team?
 - Is your role to provide treatment targeting functional activities?
- Diagnostic Assessment- use of impairment-based, standardized tests are helpful. These types of tests are NOT helpful to form a functional plan of care. They also won't typically capture functional gains in this population.
- Person-Centered Assessment: Life Participation Approach
 - Client –directed rather than clinician-directed: individual and family members play an active role in identifying therapy goals
 - Focuses upon utilizing individual's preserved strengths, rather than identifying and strengthening weaknesses
 - All goals focus upon meaningful, purposeful activities; integrating use of strategies into individual's daily routines and activities
 - Actively engaging care partners, family members, and care givers into assessment and treatment sessions, in order to promote generalization of strategies
 - Standardized testing may be used to further investigate cognitive areas that are impacting life participation
 - Utilize standardized tests that focus upon participation in functional activities, such as the Assessment for Living with Aphasia (ALA), Assessment of Language-Related Functional Activities (ALFA), Communication Activities of Daily Living (CADL-3), Communication Confidence Rating Scale (CCRSA), or Social Networks Inventory (SNI)
 - Completing education and training throughout the evaluation
 - Determining what strategies will need to be adjusted over time and introducing new strategies that will be needed in the next phase of the disease

Goal Writing for People with PPA

- Developing plan of care should always include the input from the individual and family
- Goals should be individualized and should focus upon functional outcomes
- Goals should be realistic; may need adjust the level of cueing as the disease progresses so that goals can be met (in order to ensure reimbursement)

Home Exercises:

- Rehearsal of personally relevant words: home program designed to increase person's word retrieval and pronunciation of words that they frequently use during daily conversation

- Strategies to help with pronunciation of multisyllabic words: watching a care partners lips while they say the target word together; care partner slowly fades out while person continues to say the word, writing the target words on flashcards and drawing lines between each syllable, singing or tapping the syllables out on target words
- Script Training: person with aphasia and caregiver are trained to type up and rehearse scripts, to increase ability to speak via telephone, tell stories or jokes, order food at a restaurant, say prayers, or explain their condition to others; For those with pronunciation difficulty: record video of script, of a mouth producing the script and over-articulating each word. The person chorus-reads the script along with the video. Scripts need to be practiced intensively (5-10x in a row, multiple days each week)
- Copy & Recall Treatment (CART): Written rehearsal of personally relevant words, to facilitate word retrieval or written communication of the target word
- Personal Picture Description Task: person with aphasia selects favorite photographs from present or past, and described them in 3-5 sentences. Key words/phrases are written on the back of the pictures to facilitate word retrieval as needed

Compensatory Strategies for Communication, Reading and Writing:

- Self-cueing and nonverbal communication strategies: semantic circumlocution (talking around the word), phonemic self-cueing (thinking of first letter), writing, gestures, visualization
- Communication Partner Training: on positive communication strategies at the conversation level (e.g., effectively cueing for increased use of semantic circumlocution by prompting “Tell me about it.” or whether or not to fill in the word for the person with aphasia, based upon the person’s preference)
- Facilitating auditory comprehension: training caregiver on strategies such as simplifying sentence structure, slowing rate or increasing use of nonverbal cues (write out key words during conversation or show a picture to help with comprehension)
- Low-tech communication aids: (emphasize to person and family that they need to have a “toolbox” full of different tools to help with communication; many different types of strategies and aids to help facilitate communication)
 - Communication books or boards (word or picture-based)
 - Communication wallet: (word or picture-based) portable aid so that person can easily look up important words or information during conversation
- High-tech communication aids – need to consider:
 - Person’s motivation to use an AAC device
 - Person’s fine motor and sequencing skills (Can he successfully operate a computer, TV remote or cell phone?)

- Person's level of family support, as someone will be needed to help patient program and effectively use the device as the disease progresses
- Out of pocket cost for the person/family
- Trialing the use of a device in the person's home environment during functional situations
- Writing Strategies: Use of technology or written aids/templates to facilitate spelling and grammar.
 - Spell/grammar check on phone, tablet or computer
 - Voice Recognition Technology – to state word and have device spell word out
 - Dictionary.com or other similar websites/apps
 - Templates/written aids for writing emails, To Do lists, grocery lists, or any other functional writing task
- Reading Strategies: Use of technology and compensatory strategies to facilitate functional reading tasks
 - Novels: try listening to book on tape while following along with hard copy of novel
 - Pick novels you have read before, so you are familiar with the plot and characters
 - Pick novels that are shorter with less complex plots and fewer characters
 - Single Word Reading Comprehension: Highlight the word on your phone, tablet, or computer and select “define” OR type in word into Google search and it will display image and definition
 - Lingraphica NewsPath – free program that offers several news articles each day. Program will read content aloud, highlight words that are displayed in large font

Use of Apps for People with PPA

- Train as early as possible in the disease and always complement with simpler strategies not involving technology, such as a communication wallet/book (Holland et al., 2012):
 - a. Story Creator –photo album app: can be used for communication purposes, such as telling stories
 - b. Pic Collage – Aid Making app: can easily make personalized communication boards or books
 - c. Microsoft PowerPoint – Aid Making app: can easily make communication wallets (portable aids) with this program. One slide = one page of wallet. Print 4 or 6 slides to a page, then cut out, laminate and place on a key ring
 - d. vBox – reading app: will read emails aloud to user

- e. FlipCards Pro – quiz app: allows user to make flashcards with pictures for word rehearsal.
- f. Quizlet – quiz app: allows user to make flashcards with automatic auditory cues. Can add pictures if you pay small fee for the app.
- g. Scene&Heard – AAC app: easy to navigate, uses person’s own pictures in different scenes
- h. Locabulary – AAC app: user must be able to construct sentences when given choices
- i. Dynavox Compass – AAC app: communication app, using own pictures to create scenes and scripts

Psychosocial Considerations

Conditions such as Primary Progressive Aphasia present many challenges for individuals and their family members. Due to the impairment of language and the progressive nature of the condition, capacity for relational connection is altered in varied ways and can affect couple bonds and family life. Roles and responsibilities inevitably change. Caregiving stresses over time can have serious health and mental health consequences and reverberate throughout the family and other relationships. Key to adapting and coping is a reliance on the strengths and abilities of the diagnosed person, care partners and the family as a whole.

Responses to a diagnosis of PPA

The way person and their family react to a diagnosis of PPA and the ways they adapt and cope are influenced by a range of factors such as their personality, their previous experiences, their understanding of PPA, the social and emotional support they receive and their environment. People will react differently and may adopt different strategies at different times. Many individuals with PPA are aware and insightful regarding their diagnosis and changes in language and communication; however, some may not (or be able to) acknowledge that things are becoming more difficult.

Identity

A person’s sense of identity is shaped by many things, including their relationships, roles in the family and community, hobbies and occupation. Changes in people’s abilities caused by PPA, however, may lead to changes in their sense of identity. It is important that the people around the person with PPA are aware of this as they are able to influence how the person sees themselves. Treat the person with PPA as an individual and value them as such rather than defining them by the condition or focusing on negative aspects such as lost abilities.

Relationships, roles and responsibilities

Relationships with the people around us form a central part of our identity. Relationships can change when someone has PPA. People with PPA can easily become isolated or avoided by those around them. They may lose contact with friends and family, who may fear not knowing how to react to and communicate with them. It is important to support existing relationships and encourage continued participation in social groups, community activities, religious activities and hobbies. The progressive changes in language and other areas of functioning due to the diagnosis of PPA may also change the relationships between the person and those closest to them, who may find they have taken on the identity of a 'caregiver'. A caregiver may find they have an increasing number of roles in the relationship and feel like they are on an emotional roller coaster. Caregivers are at increased risk for depression. On the one hand, caregiving can also be a rewarding personal experience. On the other hand, exhaustion, worry, inadequate resources and continuous care demands are enormously stressful. Paying attention to and helping caregivers take responsibility for their own self care is part of good patient care overall.

Options for Information and Support

PPA Education

- Northwestern University Cognitive Neurology and Alzheimer's disease Center <http://www.brain.northwestern.edu/dementia/ppa/index.html>
- Association for Frontotemporal Degeneration <http://www.theaftd.org>
- National Aphasia Association <https://www.aphasia.org>

Legal and Financial Resources

- Social Security Administration <https://www.ssa.gov>
- National Academy of Elderlaw Attorneys <http://www.naela.org>

Respite Care and Support

- Administration for Community Living <https://www.acl.gov>
- Home Care Association of America <http://www.hcaoa.org>
- Adult Day Services <https://www.nadsa.org>

Contact us for information on research opportunities, clinical care, and education opportunities:

brain.northwestern.edu | communicationbridge@northwestern.edu | 312-503-4012

References:

- Beeson, P.M., King, R. M., Bonakdarpour, B., Henry, M.L., Cho, H & Rapcsak, S.Z (2011). Positive effects of language treatment for the logopenic variant of primary progressive aphasia. *J Mol Neurosci*, 45(3), 724-736.
- Croot, K., Nickels, L., Laurence, F., & Manning, M. (2009). Impairment- and activity/participation-directed interventions in progressive language impairment: Clinical and theoretical issues. *Aphasiology*, 23(2), 125-160.
- Decker, D. A., & Heilman, K.M. (2008) Steroid treatment of primary progressive aphasia. *Arch Neurol*, 65 (11), 1533-1535.
- Farrajota, L., Maruta C, Maroco J, Martins I.P, Guerreiro M., & de Mendonça A (2012) Speech therapy in primary progressive aphasia: a pilot study. *Dementia and Geriatric Cognitive Disorders*, 2:321–331
- Finocchiaro, C., Maimone, M., Brighina, F., Piccoli, T., Giglia, G., & Fierro, B. (2006). A case study of primary progressive aphasia: Improvement on verbs after rTMS treatment. *Neurocase*, 12, 317-321.
- Henry, M.L., Meese, M.V., Truong, S., Babiak, M.C., Miller, B.L., & Gorno-Tempini, M.L. (2013). Treatment for Apraxia of Speech in Nonfluent Variant Primary Progressive Aphasia. *Behavioral Neurology*, 26(1-2), 77-88.
- Holland, A.L., Weinberg, P., & Dittelman, J. (2012). How to Use Apps Clinically in the Treatment of Aphasia. *Seminars in Speech and language: Adult focus*, 33 (3), 223-233
- Johnson, N.A., Rademaker, A., Weintraub, S., Gitelman, D., Wienecke, C., & Mesulam, M. (2010). Pilot trial of memantine in primary progressive aphasia. *Alzheimer Dis Assoc Disord*, 24(3), 308.
- Kertesz, A., Morlog, D., Light, M., Blair, M., Davidson, W., Jesso, S., & Brashear, R. (2008). Galantamine in frontotemporal dementia and primary progressive aphasia. *Dement Geriatr Cogn Disord*, 25(2), 178-185.
- McNeil, M.R., Small, S.L., Masterson, R.J., & Tepanta, R.D. (1995). Behavioural and pharmacological treatment of lexical-semantic deficits in a single patient with primary progressive aphasia. *American Journal of Speech-Language Pathology*, 4, 76-93.
- Reed, D.A., Johnson, N.A., Thompson, C., Weintraub, S., & Mesulam, M-M. (2004). A clinical trial of bromocriptine for treatment of primary progressive aphasia. *Annals of Neurology*, 56(5), 750.
- Sapolsky, D., Bakkour, A., Negreira, A., Nalipinski, P., Weintraub, S., Mesulam, M.-M., Caplan, D., & Dickerson, B.C. (2010). Cortical neuroanatomic correlates of symptom severity in primary progressive aphasia. *Neurology*, 75 (4), 358-366.
- Youmans, G. L., Holland, A. L., Munoz, M. & Bourgeois, M. (2005). Script training and automaticity in two individuals with aphasia. *Aphasiology*, 19, 435-450.