

Nonfluent Primary Progressive Aphasia

Implications of Palliative Care Principles for Informing Service Delivery

Nidhi Mabendra and Ashlyn Tadokoro

Purpose: Nonfluent primary progressive aphasia (nfvPPA) is an established language-led dementia and a known variant of frontotemporal degeneration. The purpose of this article is to report the trajectory of a single case, AC, diagnosed with nfvPPA. We describe a range of interventions offered to AC over 3 years that aimed to support her communicative function, social participation, sense of identity, and emotional well-being. We offer reflections on how these interventions align with palliative care principles, highlighting their value for guiding communication and life participation interventions for persons with nfvPPA. **Method:** This study used a longitudinal analysis of impairment progression, traditional outcome measures, patient-reported outcome measures, and narrative description to report on patient and family response to interventions. **Results:** Individual and group interventions are critical for addressing the progressive communication impairments and life participation restrictions for persons affected by nfvPPA. Palliative care principles have intuitive appeal for informing the selection of intervention approaches while focusing on managing symptoms, upholding patient autonomy and dignity, and enhancing quality of life throughout the course of rapidly progressive conditions like primary progressive aphasia. **Discussion/Conclusion:** Speech-language pathologist-led interventions, grounded in palliative care principles, can optimize communicative function and quality of life for persons with nfvPPA. **Key words:** *communication, interventions, palliative care, primary progressive aphasia, quality of life*

PALLIATIVE CARE is a specialized branch of medicine that is devoted to optimizing the health and well-being of any person

with a serious, long-term health condition. The focus of palliative services is to minimize undue suffering, uphold client autonomy and dignity, and to optimize client and family quality of life (QoL) against the odds of a life-limiting and capacity-limiting diagnosis (World Health Organization, 2003). Palliative care frameworks are of particular importance in informing service delivery for persons with neurodegenerative disorders that lack a cure and are accompanied by inevitable, progressive decline of cognitive-communicative function. Palliative care models provide a more aspirational standard for service provision than the eschewed therapeutic nihilism that results in severely restricted services or no services for persons affected by such conditions. Therapeutic nihilism can deter clinicians due to skepticism or undue

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pessimism about the value of communication interventions for clients with progressive neurodegenerative conditions (Dunkelman & Dressel, 1994). Further, a palliative care framework seems especially relevant in the person-centered care of persons with neurodegenerative conditions because clinicians can play an important role in supporting persons with neurodegenerative conditions through “letting go” (Vescovich, 2015) of daily cognitive-communicative functions (speaking, select activities of daily living, and life roles). Primary progressive aphasias (PPAs) or language-led dementias are an example of one such neurodegenerative condition that benefit from a palliative framework that aims not to restore or improve communicative function, rather to ensure communicative access for affected persons throughout the disease course. To better understand PPA, we begin with a discussion of the link between progressive aphasias and frontotemporal degeneration (FTD).

Frontotemporal degeneration represents a heterogeneous group of brain disorders, resulting from progressive degeneration of the frontal and/or temporal lobes of the brain. Among persons younger than 60 years, FTD is the most common dementia and affects approximately 60,000 Americans (The Association for Frontotemporal Degeneration, 2019). Being a rare condition, many medical providers and rehabilitation clinicians are unfamiliar with FTD, and this low awareness of FTD typically results in a 3- to 4-year delay in accurately diagnosing and identifying an affected person’s particular FTD variant (van Vliet et al., 2013). Frontotemporal degeneration has multiple variants—a behavioral variant (bvFTD), three language variants or PPAs, and known motor variants (e.g., corticobasal syndrome and progressive supranuclear palsy). Among the PPAs are a nonfluent or agrammatic variant (nfvPPA), a semantic variant (svPPA), and a logopenic variant (lvPPA). Primary progressive aphasia was first described by Mesulam (1982) and is characterized by gradual, pro-

gressive loss of language and communicative function with relative sparing of other cognitive domains and activities of daily living in the first 2 years of the disease (Gorno-Tempini et al., 2011; Mesulam, 2001). Much after Mesulam’s initial description of PPA, Kertesz et al. (Harciarek & Kertesz, 2011; Kertesz et al., 1994) presented evidence justifying the inclusion of PPA as a type of FTD. Since then, considerable research on FTD and PPA has resulted in a clearer understanding of the clinical phenotypes of PPA, their neurologic, pathophysiologic, and genetic bases as well as distinctive profiles of speech, language, cognition, and functional communication (Gorno-Tempini et al., 2004, 2008, 2011; Henry et al., 2018).

Currently, there is no cure or pharmacological treatment for PPA (or FTD) and the primary approach to care is palliative in nature to maintain and enhance QoL (Douglas, 2014; Volkmer et al., 2019) in the face of deteriorating communicative and cognitive function. Indeed, some experts have suggested “the best we can do is counsel, support, and help the families of patients with this devastating collection of diseases” (Hodges & Piguet, 2018). Yet there is a growing evidence base that reveals the value of a phased treatment approach by speech-language pathologists (SLPs; Hinshelwood et al., 2016) utilizing varied compensatory approaches for managing the cognitive-communicative challenges resulting from PPA (Dial et al., 2019; Tippet et al., 2015; Volkmer et al., 2019). Further, life participation approaches (Chapey et al., 2000; Rogalski & Khayum, 2018), lifestyle interventions employing music and choir participation (Fogg-Rogers et al., 2016; Tamplin et al., 2013; van der Steen et al., 2018), art (Chancellor et al., 2014; Deshmukh et al., 2018) and regular physical activity (Law et al., 2020) also are emerging as having an important role in modulating cognitive-communicative changes and enhancing mood and overall well-being.

Experts concur that FTD and PPA are particularly devastating for younger persons who often experience delayed diagnosis and are

typically diagnosed in the prime of their lives, while actively engaged in occupational, parenting, and caregiving roles. Volkmer et al. (2018) have reported that many medical providers do not refer persons with PPA to SLPs because they are not knowledgeable about the benefits of speech and language therapy and functional communication interventions for PPA. Given the progressive nature of PPA, the prompt timing and careful selection of palliative interventions for supporting communication is critical for patients and families. In thinking about palliative interventions, it is useful to consider some basic principles of palliative care. In the American College of Surgeons (2005) identified 10 principles of palliative care (Table 1) to guide practitioners providing care to persons with serious health conditions.

These principles have important implications for SLPs who have a vital role in providing clinical services to persons with PPA. These principles articulate the inherent value of person-centered and family-centered care; preserving patient autonomy and dignity; the role of clinicians in supporting the well-being of patients and families by reducing undue disease burden; thoughtful collaboration on goals of care with patients, families, and members of the health care team. Clinicians have to balance the limited (albeit growing) evidence available for informing them about direct and indirect interventions that may be beneficial for persons with PPA, with patient and family preferences, and clinicians' ethical duty to ensure communicative access and participation in holistic interventions for preventing depression and improving patient and family QoL. With this background, the focus of this article is on reporting the trajectory of a single case, AC (fictitious initials), diagnosed with the nonfluent variant of PPA (nfvPPA). We describe a range of interventions that were offered to AC over 3 years that aimed to support her communicative function, social participation, sense of identity, and emotional well-being. We offer reflections on how these interventions aligned with the aforementioned palliative care

Table 1. Principles of palliative care

| | |
|--------------|--|
| Principle 1 | Respect the dignity and autonomy of patients and their family caregivers. |
| Principle 2 | Honor the right of a competent person to participate in establishing goals of care and choosing among treatments. |
| Principle 3 | Communicate effectively and empathetically with patients and their families. |
| Principle 4 | Identify primary goals of care from the patient's perspective, and address how clinical care can achieve these objectives. |
| Principle 5 | Strive to alleviate pain and other burdensome physical and nonphysical symptoms. |
| Principle 6 | Recognize, assess, discuss, and offer access to services for psychological, social, and spiritual issues. |
| Principle 7 | Provide access to therapeutic support that can realistically improve patient-perceived quality of life, throughout the disease trajectory. |
| Principle 8 | Recognize physician and clinician responsibility to discourage treatments that are unlikely to achieve patient goals. |
| Principle 9 | Arrange for continuity of care by physicians, alleviating the sense of abandonment patients experience when "curative" therapies are no longer useful. |
| Principle 10 | Maintain a collegial and supportive attitude toward others entrusted with patient care. |

principles, highlighting their value throughout the article for informing the choice of speech, language, and life participation interventions for persons with nfvPPA. Next, we discuss the characteristics of nfvPPA.

The nonfluent or agrammatic variant of PPA is characterized by early appearance of syntactic errors in spontaneous speech, progressing reduction in verbal output, impaired repetition, impaired syntactic processing (e.g., reduced ability to conjugate verbs and prepositional errors), caregiver report of deteriorating verbal communication, and gradual impairments of social cognition (Douglas, 2014; Hodges & Piguet, 2018). Nestor and Hodges (2000) appropriately described the language production impairment in nfvPPA as “Broca-like aphasia” (p. 441) that eventually progresses to a state of mutism. Also, it is common for nfvPPA to coexist with progressive apraxia of speech (AoS) or a combination of AoS and dysarthria (Ogar et al., 2007). As nfvPPA progresses, it also affects the ability to read aloud and silently (Brambati et al., 2009) as well as spelling ability (Sepelyak, et al., 2011). Further, recent research by Hardy et al. (2019) has revealed that many persons with nfvPPA present with impaired hearing and other auditory symptoms such as understanding speech-in-noise, and auditory recognition of voices and environmental sounds. Neuroimaging evidence reveals that nfvPPA is marked by left frontal and left anterior insula atrophy (Wittenberg et al., 2008), as well as recent evidence of more defined atrophy of the left superior precentral region of the dorsal anterior insula (Mandelli et al., 2016).

METHODS

Case description: Background and referral history

At the time of initial referral, AC was a college-educated, married, 57-year old, Caucasian woman of Italian descent. She had 2 adult children and was a physically fit, active person who ran her own successful cater-

ing business. Her medical history was significant for postmenopausal hormone replacement therapy, hypothyroidism, insomnia, intermittent depression, and bilateral moderate hearing loss with onset in adulthood due to exposure to recreational firearms. She had been prescribed hearing aids and wore these inconsistently. AC also was an avid reader, talented chef, and enjoyed visual art. The earliest clinical episode reported by AC took place 5 years ago after a yoga class. AC woke up with a severe headache and stiff, painful neck necessitating a visit to the local emergency room (ER), where she was given a complete workup for meningitis including a computerized tomography (CT) scan and lumbar puncture. Results of this workup were negative for meningitis.

In AC’s recollection and as corroborated by her husband and daughter, she began to notice gradual speech and language changes over the 2 years subsequent to this ER visit. She described these changes as a “speech impediment” with effortful speech and errors, omission of verbs, and overuse of stereotypical utterances (e.g., “That’s great”), as well as errors in using the correct pronoun (e.g., “he” for “she”) and correct tense (e.g., “visited” for “will visit”). During this time, family members described AC as catching her errors spontaneously and being aware that she had made an error. The family’s concern peaked after witnessing AC struggling to make congratulatory remarks at a family celebration. AC also recalled becoming more conscious of her difficulty speaking and asking close friends if they noticed any changes in her speech. Per her self-report, her friends reassured her that they did not notice changes. It was her daughter who affirmed that AC’s speech was “not right” and had changed. AC also described feeling at this time that her mind was “slowing down.”

Subsequently, AC and her family were referred by their primary care physician (a family medicine specialist) to a specialty neurology clinic where she was eventually diagnosed as having PPA based on results from a detailed client and family interview, clinical

examination, laboratory work, MRI scans, and neuropsychological testing. Her MRI scans revealed a left insular opercular atrophy on axial fluid-attenuated inversion recovery (FLAIR) imaging and diffusion-weighted imaging with minimal periventricular white matter disease (on FLAIR imaging), and no atrophy of the cerebellum or the bilateral hippocampi, thus providing an imaging-supported diagnosis of FTD and being consistent with a diagnosis of PPA. Clinical testing revealed that her particular speech and language impairments were consistent with *nvPPA*, as specified by Gorno-Tempini et al. (2011). Specifically, she presented with agrammatism, morphosyntactic errors, apraxia of speech, impaired comprehension of grammatically complex sentences while having spared single-word comprehension and spared semantic knowledge. Following this diagnosis, AC was advised by the neurologist overseeing her workup to increase her physical activity and to exercise daily, to remain socially active and intellectually stimulated, and to be watchful for onset of signs of depression. Given that this specialty clinic was located more than a 2-hour drive from AC's residence and was not connected via public transit, she was advised to seek a local neurologist to oversee her care and directly referred her to our university-based clinic for speech-language pathology services. AC came to our clinic about 3 months after her formal diagnosis.

Approach to assessment

AC received a comprehensive speech and language evaluation at our clinic, 3 years ago with annual reevaluations of her cognitive-communicative functioning. The goal of this initial evaluation was to establish baseline cognitive-communicative function prior to implementing impairment-based or life participation interventions, to identify impaired versus spared cognitive abilities, and to use evaluation data to inform our choice for interventions (Mahendra & Hopper, 2016). Consistent with palliative care Principles 1 and 3 (Table 1) highlighting patient autonomy and compassionate communication about a

progressive condition, and Principles 2 and 4 focusing on collaborative goal-setting, we wanted to use assessment data to educate and counsel AC and her family, predict skills that were likely vulnerable to future decline, and develop meaningful goals of care for AC and her family.

We used the Western Aphasia Battery-Revised (WAB-R; Kertesz, 2006) as a standardized, impairment-based measure to track the progression of her cognitive-communicative impairments over time. We chose the WAB-R also knowing that it allows for assessment of nonverbal cognition (using Block Design and Raven's Progressive Matrices), apraxia, reading, and writing performance. We supplemented this standardized assessment with regular client and family member interviews, brief assessments of mental status (i.e., Mini-Mental State Examination at initial evaluation; the Montreal Cognitive Assessment subsequently), and nonstandardized testing using select subtests of other batteries, as needed. Table 2 presents AC's performance on the WAB-R over time. Given the emphasis placed on using patient-reported outcome measures (PROMs; Irwin, 2012) in person-centered assessment and the critical importance of monitoring the emotional well-being of a person with aphasia (Worrall, 2019), we wanted to monitor AC's self-reported QoL. The use of PROMs and QoL measures also aligns with palliative care principles (Table 1) that emphasize providing therapeutic support to improve patient QoL (Principle 7) by identifying and addressing burdensome symptoms (Principle 5), social participation, and psychological issues (Principle 6). To this end, we used an intake interview at initial evaluation, the American Speech-Language-Hearing Association (ASHA) Quality of Communication Life Scale (QCLS; Paul et al., 2004) in Years 1 and 2, and the Aphasia Impact Questionnaire (AIQ; Swinburn et al., 2018) in Year 3 to capture AC's unique perspectives on her rapidly changing speech and communication ability. In Year 3, we changed our choice of measure from the ASHA QCLS to the more recently developed AIQ given its greater depth and

Table 2. AC's performance on the Western Aphasia Battery-Revised

| Initial Evaluation WAB-R | Year 1 WAB-R | Year 2 WAB-R | Year 3 WAB-R Bedside ^a |
|---|--|--|---|
| <i>Spontaneous speech</i> 14/20 Content = 9/10 Fluency = 5/10 | <i>Spontaneous speech</i> 13/20 Content = 9/10 Fluency = 4/10 | <i>Spontaneous speech</i> 7/20 Content = 5/10 Fluency = 2/10 | <i>Spontaneous speech</i> Content = 1/10 Fluency = 0/10 |
| <i>Auditory verbal comprehension</i> 9.65/10 | <i>Auditory verbal comprehension</i> 7.3/10 | <i>Auditory verbal comprehension</i> 4/10 | <i>Auditory verbal comprehension</i> 3/10 |
| Yes/no questions 57/60 Word recognition 60/60 Sequential commands 76/80 | Yes/no questions 57/60 Word recognition 56/60 Sequential commands 33/80 | Yes/no questions 40/60 Word recognition 30/60 Sequential commands 10/80 | |
| <i>Repetition</i> 6.4/10 <i>Naming</i> 8.1/10 Object naming 47/60 Word fluency 14/20 Sentence completion 10/10 Responsive speech 10/10 | <i>Repetition</i> 2.7/10 <i>Naming</i> 3.4/10 Object naming 17/60 Word fluency 10/20 Sentence completion 2/10 Responsive speech 5/10 | <i>Repetition</i> 1/10 <i>Naming</i> 1.2/10 Object naming 8/60 Word fluency 2/20 Sentence completion 0/10 Responsive speech 2/10 | <i>Repetition</i> 0/10 <i>Naming</i> 0/10 |
| <i>Apraxia quotient</i> 76.4 <i>Drawing</i> 30/30 | <i>Apraxia quotient</i> 52.8 <i>Drawing</i> 28/30 | <i>Apraxia quotient</i> 26.4 <i>Drawing</i> 26/30 | Bedside aphasia score 5 Bedside language score 13.75 |
| <i>Block design</i> 9/9 <i>Raven's matrices</i> 37/37 | <i>Block design</i> 9/9 <i>Raven's matrices</i> 37/37 <i>Apraxia screening</i> 47/60 <i>Reading score</i> 16/20 <i>Writing score</i> 16/20 | <i>Block design</i> 9/9 <i>Raven's matrices</i> 35/37 <i>Apraxia screening</i> 39/60 <i>Reading score</i> 10.2/20 <i>Writing score</i> 16/20 | <i>Block design</i> 6/9 <i>Raven's matrices</i> 30/37 <i>Apraxia (bedside)</i> 6/10 <i>Reading</i> Unable <i>Writing</i> 8/10 |

Note. WAB-R = Western Aphasia Battery-Revised.
^aAdministering the full WAB-R was no longer appropriate for AC at the end of Year 3; instead, we used the WAB-R Bedside and select WAB-R Part 2 subtests to track her performance.

focus on assessing the impact of living and coping with aphasia, specifically across the domains of emotional well-being, functional communication, and social participation. Further, the AIQ fits better with palliative care Principle 6 (Table 1) that highlights the need to assess, discuss, and consider services for psychological issues (i.e., emotional states and view about the future) and social participation (i.e., loneliness, social isolation, and positive activities).

Table 3 shows results of AC's longitudinal performance on cognitive status measures, PROMs, and other tasks. Finally, Table 4 presents a narrative summary of how AC's language and communicative function changed as her PPA severity progressed. These data in Tables 2, 3, and 4 reveal the relentless, longitudinal progression of impairments associated with nvfPPA. These data offer strong impetus for continually improving the accuracy of early identification of PPA and early referral to SLPs.

Intervention recommendations

Per guidelines published by ASHA (ASHA, 2005a, 2005b), the roles and responsibilities of SLPs working with persons with a progressive neurodegenerative condition like PPA include (a) assessment of cognitive-communicative functioning, (b) collaboration with clients and families to develop meaningful intervention plans, (c) education and counseling of clients, families, and caregivers about impairment progression, (d) client advocacy and assisting with case management, and (e) research to better understand effects on cognitive-communicative function and to develop evidence-based interventions. With these roles in mind for SLPs, palliative care principles for persons with progressive, capacity-limiting conditions (Table 1), and AC's assessment data (Tables 2 and 3), her goals of care differed as time went on and intervention typically included individual and group treatment. We describe our phased approach to interventions with AC, detailed next.

Year 1: Diagnosis of nvfPPA

Individual interventions

An early theme for AC and her family was their immediate need to fully understand her diagnosis and its implications, to address relevant comorbidities, and to process the grief resulting from her diagnosis. In thinking back to palliative care principles, we wanted to uphold AC and her family's autonomy (Principle 1), invite their input in selecting initial goals of care (Principle 2), strive to alleviate pain, grief, and other symptoms (Principle 5), and provide quick access to therapeutic supports (Principle 7). We began with educating AC and her family about the nature and trajectory of nvfPPA and its relationship to FTD, discussing AC's spared and more impaired abilities, and counseling and advising the family to put their financial and business affairs in order (because AC was the owner and sole operator of a business). We addressed related comorbidities of AC's hearing loss and chronic, disrupted sleep via referrals to an audiologist and to a specialist in sleep medicine. We discussed with AC her candidacy for SLP interventions to help enhance and maintain functional communication, and also provided assistance and resources in locating relevant clinical trials for which AC might be a candidate. AC and her family elected not to pursue pharmacological intervention trials. AC expressed strong interest in wanting to preserve spoken communication and her deep desire to have access to a larger community of other adults coping with aphasia or PPA.

Given her fairly mild impairments at initial presentation accompanied by high motivation, our approach to AC's therapy in the first year (one 12-week session of therapy per semester and a 6-week therapy session in the summer) focused on individual and group interventions. In individual therapy, AC was seen twice weekly to address client and family education and counseling—communication partner training and conversational coaching using strategies for Supported Conversation for Aphasia (SCA; Kagan et al., 2001) and evidence-based strategies for

Table 3. AC's performance on other cognitive-linguistic and QoL measures

| Initial Evaluation | Year 1 | Year 2 | Year 3 |
|---|---|--|---|
| <i>Cognitive status</i> Mini-Mental State Examination 28/30 No evidence of global cognitive decline. | <i>Cognitive status</i> Montreal Cognitive Assessment (MoCA) 21/30 <i>ASHA Quality of Communication Life Scale</i> AC reported QoL of 3 on a 5-point scale (1 = low, 5 = high) | <i>Cognitive status</i> MoCA 17/30 <i>ASHA Quality of Communication Life Scale</i> AC reported QoL of 4 on a 5-point scale (1 = low, 5 = high) | <i>Cognitive status</i> No MoCA testing <i>Aphasia Impact Questionnaire (AIQ-21)</i> AC reported feeling positive about the future, confident, and reported having positive things to do. AC also expressed being socially isolated and bored. |
| <i>Other measures</i> <i>Arizona Battery for Communication Disorders of Dementia (ABCD)-Conceptual Knowledge</i> Concept Definition Subtest 46/60 <i>Texas Functional Living Scale</i> Communication Subscale Score 21/28 | <i>Other measures</i> <i>WAB-R Written Picture Description</i> Five telegraphic sentences with nouns, verbs, and conjunctions. e.g., "Dad and mom reading and drinking wine," "Family am happy." | <i>Other measures</i> <i>WAB-R Written Picture Description</i> Six telegraphic sentences with nouns and verbs, no conjunctions. e.g., "Daughter bring sand balance," "Father and mom is read book and wine drink," "Son am kite, dog." <i>Sample written discourse task</i> Weekend activity description (Figure 1) | <i>Other measures</i> <i>WAB-R Written Picture Description</i> Three telegraphic sentences with nouns and verbs, no conjunctions. e.g., "Dad and mom, wine, drink," "Son am kite, dog." <i>Sample written discourse tasks</i> (Figure 2) <i>Sample language/art expression task</i> (Figure 3) |

Note. ASHA = American Speech-Language-Hearing Association; QoL= quality of life; WAB-R = Western Aphasia Battery-Revised. Administering the MoCA was no longer appropriate for AC at the end of Year 3, given its reliance on verbal responses.

Table 4. AC's changing communication skills

| Feature | Initial Evaluation | Year 1 | Year 2 | Year 3 |
|--------------------------|--|--|---|---|
| Speech sound distortions | Occasional speech sound distortions and paraphasias. | Frequent speech sound distortions and phonemic paraphasias. | Very frequent speech sound distortions and phonemic paraphasias. | Nonverbal. Rare vocalization of vowels. |
| Rate of speech | Reduced speech rate. Occasional pauses. | Reduced rate of speech. Effortful, halting speech; multiple filled pauses. | Marked reduction in rate of speech. | Nonverbal. |
| Frank syntactic errors | AC's first symptoms were pronominal errors (e.g., using "she" for a man) and errors in using plural markers. | Frank syntactic errors, fewer verbs and conjunctions used. | Pervasive syntactic errors. Telegraphic output, loss of conjunctions. | Nonverbal. |
| Lexical retrieval | Impaired verbal word-finding ability. Spared conceptual knowledge. | Impaired verbal word-finding ability. Impaired conceptual knowledge of objects. | More impaired verbal word-finding ability. Retained object knowledge (in writing). | Nonverbal. Written word-finding ability spared. |
| Repetition | Impaired repetition. | Moderate impairment of repetition. | Severe to profound impairment of repetition. | Nonverbal. |
| Auditory Comprehension | Initial, mildly impaired auditory comprehension. No impairment for answering yes/no or two-choice questions. | Mild impairment of auditory comprehension, affects understanding of long, multistep or grammatically complex utterances. | Increasing auditory comprehension impairment. Unable to engage in conversation without written/pictographic supports or use of an AAC device. | Severe impairment of auditory comprehension. Requires all input to be written, visual, pictographic, or gestural. |
| Reading | Avid reader; minimal impairment of reading comprehension. | Mild impairment of reading comprehension. | Mild impairment of paragraph-level content. Moderate impairment of ability to read stories or books. | Moderate to severe impairment of reading comprehension. Spared ability to read short sentences and questions. |

(Continues)

Table 4. AC's changing communication skills (Continued)

| Feature | Initial Evaluation | Year 1 | Year 2 | Year 3 |
|-------------------------|--|---|--|--|
| Spelling | Occasional spelling errors; self-identification of errors and attempts to correct errors. | Frequent spelling errors, typically seen for longer words. | More frequent spelling errors. Spontaneous spelling in written discourse better preserved than spelling to dictation. e.g., <i>palance</i> for palace, <i>break</i> for bread, <i>crump</i> for crumb. Written output decreased in length with more syntactic errors. Yet written communication effective and functional and used frequently in social settings. | Similar to Year 2; maintained performance on spontaneous spelling in written discourse. Severely impaired spelling to dictation. |
| Writing | Mildly impaired written expression. Reduced sentence length, some syntactic errors. Spontaneously identifies errors. | Written expression maintained. Use of agrammatic sentences, yet AC independently able to write multiple, on-topic ideas on any topic. | | No further decline in written output noted. AC still able to communicate functionally, using writing. |
| Nonverbal communication | Unimpaired. Good eye contact, attentive to communication partners, uses gestures effectively. | Strong social pragmatics; appropriate and effective use of facial expression, gesture, tone of voice, and humor. | Social pragmatics sometimes inappropriate (e.g., walking away mid-conversation or mid-interaction), altered eye contact, reduced attention and responsiveness to communication partners. Effective use of touch, smiling, pointing, and gesture. | Similar nonverbal communication as previous time point, with no further decline. |

Note. AAC = augmentative and alternative communication.

supporting verbal expression. These strategies included targeting word retrieval strategies (using object function and feature descriptors, and writing) and use of traditional script training (Youmans et al., 2011) and video-implemented script training for aphasia (Henry et al., 2018) to enhance AC's ability to communicate with her siblings and extended family members. Individual therapy was steadily supported by homework tasks designed to practice components of therapy tasks. Examples of homework tasks included copy-and-recall treatment homework for practicing writing of salient vocabulary, or rehearsing salient words and phrases along with clinician videos for practice (i.e., watch me and say with me). Similar to the findings of Henry et al. (2018), AC clearly benefited from script training, and maintained her ability to use the scripts to engage in simple face-to-face and telephone conversation with her sister and a close friend over 9 months of this first year. During this time, we observed that AC was demonstrating frequent binary reversals (e.g., confusing yes for no, she for he, and up for down) that were making communication challenging and were similar to those reported by Sivasathiaseelan et al. (2019) in three of their participants with nvPPA.

Group interventions

Given AC's desire to meet and interact with others with aphasia, we recommended that, alongside individual therapy, AC join a weekly aphasia choir and an additional supported conversation group. In the conversation group, AC benefited notably from making friends with other aphasia group members and from having information presented in multiple modalities with multiple acceptable ways of responding. She also particularly enjoyed the aphasia choir, having no difficulty with singing aloud and often reporting that music and singing put her in a better mood.

By the end of this first year, AC's spontaneous spoken output had decreased, causing her and her family much disappointment, even grief. This was used as an opportunity for active listening, empathy, and compassion (palliative care Principle 3) and

further clarifying the progressive nature of PPA, the expected nature of this decline in spoken communication, and review of what was working in individual and group therapy, and the need for augmentative and alternative communication (AAC) approaches. AC did not own an iPad and her family was advised about the benefits of acquiring an iPad so that training and use of aphasia therapy and AAC apps could be incorporated into her ongoing therapy. Her family was provided information about the importance of voice and message banking for potential future use in a speech-generating device, yet they expressed limited interest in this option. Consistent with palliative care Principle 6 and clinicians' role in offering access to additional needed services, the value of counseling services was discussed for all family members and local resources provided to avail this option. Of particular concern was the sadness expressed by AC's daughter and son, who expressed that they were "losing our mom."

Year 2: Declining speech and introduction of augmentative alternative communication

At the end of Year 1, AC was reassessed (Tables 2 and 3) to track changes in speech and language function. In consideration of AC's autonomy and right to choose among treatment approaches (Principles 1 and 2), we also wanted to obtain her perspective on treatments that were not meeting her goals (Principle 8). We used an aphasia-friendly interview (using typed questions with pictographs) to elicit AC's perceptions about what interventions had worked, what had not worked, and her preference for specific therapy goals. AC expressed a desire for more cognitive stimulation and for her therapy goals to be focused on her wish to remain actively engaged in her catering business. She expressed wanting to retain her written vocabulary and knowledge of her trademark recipes and ingredients. She communicated clearly that individual therapy and aphasia choir had been meaningful for her, and she wished to continue these interventions. She shared that she did not wish to participate

in aphasia conversation groups, as she felt increasingly challenged to speak and to understand information—we concurred with her in light of her declining ability to speak and our responsibility to use treatment time to better address her chosen goals (per palliative care Principle 8).

AC's children expressed that, given her worsening communication difficulties, they were concerned about her safety and ability to communicate when she was out in the community independently. Information was provided about MedicAlert and how to sign up for these services. In response, AC and her family obtained a medical alert ID bracelet, engraved with information about AC's diagnosis and communication difficulty, and signed up for a membership plan that offered AC access to a 24/7 emergency response team (with the team having access to her personal profile, health status information, and advance directives). Reducing the likelihood of additional burdensome physical and non-physical symptoms by ensuring AC's safety and access to prompt assistance in the event of an emergency is consistent with palliative care Principle 5 (Table 1). One challenge during this time was helping AC to transition from her long-term primary care physician to the care of an available local neurologist with experience working with persons with PPA. After much waiting, a neurologist with expertise in PPA could not be identified and AC and her family went with the first available neurologist, who did not have experience in working with persons with PPA or dementia. The family reported that this made consultations awkward, as they felt let down by their neurologist's limited understanding of PPA, FTD, and AC's needs. Further, AC had known her former physician for an extended period and was less comfortable with her new physician against the backdrop of her own declining speech.

Individual interventions

In Year 2, we continued to see AC twice weekly for individual and group therapy, during one 12-week therapy session each

semester and one 6-week summer session. In individual therapy, AC was no longer demonstrating benefit from continued script training and this was discontinued while retaining most previously prepared scripts into a communication notebook. By retaining these scripts, AC would be able to use them in conjunction with text-to-speech or handwriting-to-speech apps that would be introduced later. AC's family was again advised to participate in voice and message banking, yet chose not to pursue this option. The focus of individual sessions was to introduce and train use of an iPad (recently acquired by AC) for basic cognitive-linguistic stimulation (e.g., playing Sudoku, using the TalkPath news app for aphasia-friendly headlines, listening to music, using a calendar, and language apps involving word-to-picture matching and written naming), and for introducing AAC apps that AC could use to support her communication. Additionally, we engaged AC and her daughter in co-creating a personalized communication notebook.

This communication notebook consisted of large-font text, accompanied by pictures and pictographs to support AC's ability to communicate with family members (about her physical state, planned activities, and plans for meals) and materials to support her interactions and communicative independence at frequently visited locations (i.e., grocery store, company warehouse, and neurologist's office). Additionally, the notebook contained messages necessary for social interactions including a brief explanation of PPA and communicative strategies that are helpful for her, conversation starters for AC and for unfamiliar communication partners, as well as cherished family stories with photographs to capture major life events. The use of the communication notebook was trained and practiced during therapy sessions including role-play in conversational contexts with familiar and unfamiliar communication partners.

The iPad apps introduced to AC included Speak It (text-to-speech app for typed information, or information available on websites) and Speech Assistant AAC (a text-to-speech

communication app that allows creating phrases and categories, that can be programmed into buttons), Speak Me (an app that allows a smartphone to speak out its notifications—texts, e-mails, WhatsApp messages), and Handy Speech (an app that converts handwriting to speech). Three of these four apps worked successfully for AC, yet Speak Me was more challenging for her given her hearing loss and declining auditory verbal comprehension. Of these apps, AC used Speak It most frequently while continuing to hand-write messages on a notepad or whiteboard to communicate.

AC demonstrated continued decline in verbal communication and also was moving through her individual therapy sessions at a slower pace. Knowing time was not on our side given her PPA diagnosis and with the goal to increase ease and frequency of use of her communication notebook and AAC apps, we offered her a chance to participate in a weekly teletherapy session, via Skype. We also considered that getting AC accustomed to and comfortable with receiving therapy remotely might make it easier for her to continue to receive clinical services or have a remote communication partner. Given the growing evidence to support the feasibility and utility of teletherapy for persons with PPA (Dial et al., 2019; Volkmer et al., 2019), this seemed a viable option to plan for continuity of care for AC (Principle 9). We were pleased that, as a first-time iPad user, AC quickly became comfortable with her iPad in three training sessions, being able to log in independently for her therapy session via Skype, and responded in real time to a remote clinician by asking and answering questions via a chat box, or by using gestures (eg, thumbs up and thumbs down), holding up a whiteboard with her written response, or showing a section of her communication notebook.

Group interventions

Given AC's love of reading and relatively spared reading performance (reading score of 16/20; Table 3), we recommended that

she consider participating in an Aphasia Book Club. This recommendation was made based on our knowledge of AC's reading and written expression ability at the time, and of the use of evidence-based accommodations or reading ramps (Bernstein-Ellis & Elman, 2006) in Aphasia Book Club. AC chose to continue participating in the weekly aphasia choir and agreed to try out the Aphasia Book Club. In the first semester's 12-week therapy session, AC participated in the Aphasia Book Club for the first 6 weeks, yet was observed to have declining participation and did not benefit from reading ramps (e.g., abridged outlines, keyword/character lists, and timeline tools). She was often visibly frustrated and frequently exited book club early. We discussed this with her and mutually agreed that she stop attending Aphasia Book Club given that it was not supporting her in the goals she most valued (consistent with Principle 8 and an emphasis on discouraging treatments unlikely to achieve patient goals).

In lieu of the Aphasia Book Club, we recommended AC try a new Everyday Technology for Aphasia Care (ETAC) group at our aphasia clinic. The purpose of this group was to teach persons with aphasia practical scheduling and e-mail skills, use of WhatsApp and social media platforms, and to explore aphasia therapy apps, language games, and artistic/creative apps (e.g., for photography and collage creation). AC happily agreed to try out this group, as it appeared to be a better match for her and would allow her to use her iPad in more social contexts. One challenge during this stage was AC's occasional apathy and low motivation for participation in therapy; she usually responded well to direct counseling and messages of hope and optimism. Another challenge was the somewhat persistent belief her spouse had that the clinicians' suggestion to introduce an iPad hastened the decline of her verbal communication. This misperception was addressed in a supportive conversation about how therapy might decelerate language loss in PPA, yet cannot halt the inevitable progression

of PPA, regardless of timing of AAC device introduction.

Year 3: Progressing PPA without verbal communication

By the end of 2 years from AC's initial evaluation at our clinic, she could no longer speak any words or sentences. AC accepted this change with stoic courage and responded by writing more, and continuing to pursue art and cooking activities. Yet this was a very sad and difficult stage for her young adult children and her spouse. Milestones such as AC's 60th birthday were described by the family as deeply "touched with grief" given her total loss of speaking ability, including during laughing or crying. Family members (e.g., AC's mother, sisters, and spouse) recalled that AC had a beautiful voice and wished that they had preserved more recordings of her voice for posterity, or pursued voice banking.

Individual interventions

AC continued to participate in one, weekly individual therapy session and an additional weekly session via Skype. Individual therapy sessions remained focused on compensatory augmentative therapy using low-tech and high-tech strategies, aimed at establishing functional communication and maximizing communicative abilities in social interactions (Fried-Oken et al., 2012; Russo et al., 2017). Specifically, she wanted to maintain using her communication notebook with minimal assistance and was able to successfully retrieve biographical and other information to communicate. She also continued to use functional writing to communicate words, phrases, and short instructions salient to her catering business and for social interaction (see Figures 1 and 2 for examples of functional written communication). She was better able to use text-to-speech and handwriting-to-speech iPad apps, yet required frequent assistance.

Group interventions

Life participation-themed group interventions included weekly participation in an aphasia choir and the aforementioned func-

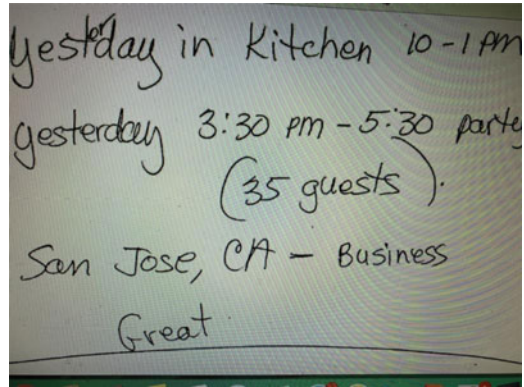


Figure 1. Year 2: Sample written response to question, "How was your weekend?"

tional technology skills group for persons with aphasia. In the aphasia choir, AC could no longer sing aloud and very rarely could hum or vocalize vowel sounds. Yet she reported that she enjoyed attending the choir even though she was primarily participating in musical activities besides the choral singing, such as vocal warm-ups (with some vocalization), and playing a kazoo. The traditional choral singing approach in the aphasia choir was adapted significantly for AC to enable her to: (1) use a kazoo, (2) use handheld musical instruments (e.g., tambourine or maracas) to keep time to the music, (3) use pictographic cues and video-modeling to create a role for her to guest-conduct the aphasia choir for specific songs using conducting gestures (e.g., pointing, hand positions for scale)

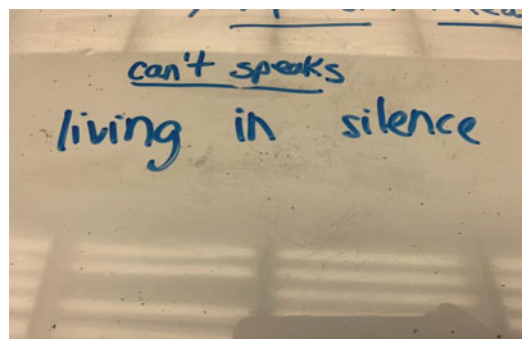


Figure 2. Year 3: Sample written response to question, "How does aphasia affect you?"

and body position (e.g., leaning or standing upright), and (4) introduce songs or aphasia advocacy messages or communicate the impact of aphasia using written communication. In the functional technology skills group, AC had much success with independent practice using therapy apps that target language abilities (i.e., spelling of single words or matching pictures to written words), memory games (targeting visuospatial and semantic memory), number games (e.g., Sudoku), and successfully created photographic collages with captions using apps like PicCollage (a photo-template design app) and VSCO (a photo-editing app). With some assistance, she also continued to send online greeting cards and to communicate with aphasia group members on a closed WhatsApp group using short, typed messages and emojis.

FUTURE DIRECTIONS FOR AC's CONTINUED CARE

In providing input about interventions that add meaning to her life, AC continues to report the value of being included in a community of people with poststroke aphasia and PPA, of participating in aphasia choir and technology skills groups, and of having an opportunity to participate in individual sessions, including teletherapy. Teletherapy likely may have rising value for AC, as she is no longer able to drive and lives in a remote area, not linked to public transit or paratransit services. She also has frequently expressed an interest in wanting to do more artistic activities (e.g., see Figure 3 for a collage she created in a therapy session to describe things especially important to her). We look to incorporate art informally into her therapy sessions while also providing a referral to a trained art therapist so AC may pursue this. Whereas AC made steady gains in using the iPad and text-to-speech apps and her writing remains quite functional for everyday communication, it is likely that she will need to transition to a more advanced, symbol-supported communication app to maintain her independence



Figure 3. Year 3: AC's creation of an identity collage, informed by Life Story Work.

in communication. Finally, AC talks openly about wanting to leave behind a chronicle of her life and work for her children and family and we hope to incorporate this goal fully into her ongoing care.

To this end, in continuing to codesign meaningful interventions with AC, we are exploring the applications of Life Story Work (Kaiser & Eley, 2017) and biographic narrative intervention (Corsten et al., 2015) to put AC's life experiences and activities into perspective through structured or unstructured therapeutic review. The goal of Life Story Work can vary from providing an individual or family reminiscence opportunity, focusing on a person's unique identity, enhancing their self-esteem and well-being, or informing a person-centered care plan. At this time, AC remains physically healthy, actively assisting in her business, and is supported by her family in continuing to attend therapy sessions.

CONCLUSION

A palliative care orientation, ultimately, empowers clinicians by asserting that interventions must change by stage of disease progression, must do no harm, and should not be tied to demonstrating improvement or even maintenance on impairment-based measures as long as interventions add meaning, value, and

QoL for a person with PPA and their family. Indeed, in adopting a palliative framework to inform the care of persons with PPA, improvement and maintenance of skills should not be required to demonstrate evidence of the value added by providing person-centered interventions. The broader issue of coping as best as possible with a progressive neurodegenerative condition is best addressed by helping patients and families find meaningful adapted or novel opportunities to participate despite diminishing skills.

It is hoped that this article provides clinicians an in-depth view of the effects of nfvPPA on cognitive-communicative functioning, as

well as the response of a single patient and their family members to varied individual and group interventions. Primary progressive aphasia has a profound impact on the communicative and cognitive ability of those affected and on their family members' social and emotional well-being. Speech-language pathologists have a vital role in making strong, positive impact on persons with PPA and their families, by initiating specific interventions to maintain and support functional communication, increase communicative support, decrease social isolation, and enhance the emotional wellness of affected persons and their families.

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