Published in final edited form as:

Int Rev Psychiatry. 2013 April ; 25(2): 237–245. doi:10.3109/09540261.2012.751017.

Behavioral Interventions for Enhancing Life Participation in behavioral variant Frontotemporal Dementia and Primary Progressive Aphasia

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Abstract

Primary progressive aphasia (PPA) and behavioral-variant frontotemporal dementia (bvFTD) are clinical syndromes under the umbrella term “frontotemporal dementia (FTD)” and are caused by a neurodegenerative disease with an onset most typically in the productive years of adulthood. The cognitive and behavioral impairments associated with FTD interfere with the successful engagement in typical life roles, such as parenting, working, and maintenance of interpersonal relationships. There are currently no treatments to stop or slow the degenerative process and there are only very limited medication options for the management of the cognitive-behavioral symptoms. However, alternative, non-pharmacological interventions may offer significant benefit to the quality of life of the diagnosed individual. The goal of this paper is to provide an overview of the approaches available through neurorehabilitation and community-based services that facilitate successful engagement in life activities and promote optimal quality of life for the individuals and families living with FTD. It is hoped that as medical providers become more familiar with behavioral interventions, referrals for services will increase thereby allowing individuals with FTD and their caregivers to learn ways to adapt, adjust, and participate in life to the fullest despite the impairments from this progressive disease.

Primary progressive aphasia (PPA) and the behavioral variant of frontotemporal dementia (bvFTD) are two clinical dementia syndromes caused by neurodegenerative brain disease. Recently published consensus criteria outline the diagnostic criteria for bvFTD and PPA (Gorno-Tempini et al., 2011; Rascovsky et al., 2011). In brief, PPA is an aphasic dementia characterized by progressive decline in language function, but relative sparing of other cognitive domains associated with damage to the left hemisphere perisylvian language network (M. M. Mesulam, 2003). Experts generally recognize three main variants of the syndrome: agrammatic (PPA-G), logopenic (PPA-L) and semantic (PPA-S), which are most conspicuous at the early stages of the disease (Gorno-Tempini, et al., 2011). The behavioral variant of FTD is a comportmental dementia characterized by change in behavior and cognition marked by features, such as apathy and disinhibition, combined with a reduced awareness about these changes (Neary et al., 1998; Rascovsky, et al., 2011) and is associated with frontal, insular and temporal atrophy. The National Alzheimer's Disease Coordinating

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Declaration of interest: The authors report no conflicts of interest.

Center (NACC) and the Uniform Data Set (UDS) of the Alzheimer's Disease Centers funded by the National Institute on Aging have adopted the diagnostic criteria for bvFTD and PPA (Morris et al., 2006). Typical age of onset for bvFTD and PPA is under age 65 and collectively they are thought to represent the most common form of young-onset dementia (Knopman, Petersen, Edland, Cha, & Rocca, 2004; Ratnavalli, Brayne, Dawson, & Hodges, 2002). While true epidemiologic data are scarce, recent consensus estimates suggest prevalence rates of FTD range between 15 and 22 per 100,000 and incidence rates are between 2.7 and 4.0 per 100,000 person-years (Knopman & Roberts, 2011).

PPA and bvFTD are clinical syndromes, not neuropathological entities. While the phenotypes and anatomic targets in clinical syndromes of PPA and bvFTD are relatively clear, the cause and their relationship to underlying pathology are less straightforward. In the majority of cases, underlying pathology of PPA and bvFTD are associated with neuropathologic changes from the frontotemporal lobar degeneration (FTLD) family including tau or ubiquitin/TDP-43 positive inclusions; however atypical Alzheimer's disease (AD) may also occur (Gefen et al., 2012; M. Mesulam et al., 2008; and for reviews see: Piguet, Hornberger, Mioshi, & Hodges, 2011; Rohrer & Schott, 2011).

There are no approved disease-modifying treatments for PPA or bvFTD that stop or slow the degeneration process. There is preliminary support for medications that may be helpful with management of the cognitive-behavioral symptoms of bvFTD and PPA. For example, antidepressants (i.e., serotonin-selected reuptake inhibitors) may improve mood and reduce some of behavioral control problems characteristic of the syndromes (for a review, see O'Brien & Burns, 2011), but the options for symptom-modifying medication options are quite limited at this point. However, there are multiple alternatives to pharmacologic intervention that may be helpful to improve quality of life for individuals with bvFTD and PPA. The current paper provides an overview of the symptoms of FTD and how behavioral interventions provide a viable option for management of symptoms and facilitation of life functioning while the individual is still living in the community. This is not meant to be an exhaustive review of the scientific literature, but rather a summary of points for medical providers to better understand the utility of behavioral interventions and the potential value for referring their patients for such interventions.

Interventions to Address the Impact of FTD on Functioning

Since disease-modifying therapies are not available and the onset of the cognitive-behavioral decline occurs during the decades of life when an individual is fully engaged in life activities, such as parenting and working, it is important to identify alternative interventions that facilitate successful engagement in life activities and promotes optimal quality of life for the individuals living with this neurodegenerative disease. Within the clinical setting, strategies should be tailored for each patient since the symptom profile and rate of decline are variable from person to person. As such, it is important to incorporate clinicians into the care plan who understand how to assess, track the impact of the symptoms on life functioning, and to apply interventions that facilitate the individual's engagement in daily life activities and reduce the burden of care as much as possible on those in the caregiver role. Individuals with FTD would likely benefit from a team-based approach to care that includes not only neurologists, nurses, neuropsychologists, social workers, and psychiatrists, but also other clinicians with expertise in behavioral interventions for addressing the symptoms from FTD that are negatively affecting life functioning. Neurorehabilitation professionals, such as Speech-Language Pathologists, Occupational Therapists, and Rehabilitation Psychologists are uniquely positioned to be of great benefit to individuals with PPA and bvFTD given their expertise in how brain dysfunction affects life functioning and how to tailor rehabilitation interventions to maximize life functioning. Individuals with

progressive dementias have not traditionally been considered a rehabilitation population because of the progressive nature of the disease process. However, that bias is changing as the field begins to apply the concepts and models of rehabilitation to address the needs of individuals with progressive dementias to participate in life activities and possibly stave off the decline as long as possible (for a review see Buchanan, Christenson, Houlihan, & Ostrom, 2011).

The field of medical rehabilitation is focused on the provision of treatments to improve an individual's functioning after the onset of injury, illness, or disease that leads to disability. Interventions generally fall into two categories, compensatory or remediative/restorative. Compensatory methods are focused on adjusting or adapting the individual's approach to life tasks or changing the environmental structure in order to facilitate compensation for the motor, sensory, and/or cognitive impairments that are present, whereas remediative/restorative interventions are aimed at improving function through addressing the underlying physiologic cause of the disability. All interventions are aimed at facilitating the individual's ability to participate in daily life activities with the lowest level of assistance as possible within the least restrictive environment.

Skill-Based Compensation Methods for Maintaining Life Functioning

Compensatory methods are aimed at adjustment or adaptation to impairments in motor, sensory, or cognitive-behavioral functioning that are present. Interventions are traditionally focused on the individual learning new skills and approaches to life tasks that reduce the reliance on the impaired area of functioning. However, for individuals with FTD, the impairments will progress over time, leading to greater impairments in life functioning. Thus, interventions are matched to the individual needs of the patient depending upon the severity of the impairments and the stage of the disease process.

Cognitive impairments

Compensation for cognitive impairments requires a comprehensive assessment of the individual's neuropsychological functioning to determine what cognitive skills are impaired. Then interventions are designed to compensate for those impaired skills so that they do not present barriers to completion of daily life tasks. The most common types of compensatory skills taught in rehabilitation are use of daily planning tools, such as a calendar or electronic device (e.g., iPad). The key for successful use of such pieces of equipment is the individual learning to plan ahead for task completion and habitually check and update the calendar and lists through-out the day. Electronic devices have the advantage over paper calendar systems of having alarm systems that can alert the individual at set times to check the schedule, take medications, or to initiate any activity, for example. Daily planning tools help the individual to compensate for impairments in memory and planning most commonly, but are even more generally helpful for assisting the individual in establishing routines that make him/her more efficient and effective for task completion. For individuals in the early stages of FTD who want to continue working as long as possible, use of such compensatory devices can be quite helpful for planning and completion of work tasks. There are a host of other compensatory skills that can be applied to help a person with FTD compensate in the early phases of the disease process for the cognitive impairments. Through the evaluation process, the neurorehabilitation professional will identify which compensatory skills are needed given the constellation of impairments. Cognitive Rehabilitation interventions are offered by Speech-Language Pathologists, Occupational Therapists, and Rehabilitation Psychologists who have expertise with working with individuals with brain injury return to community-based activities.

Communication impairments

There are two main types of communication problems that can arise with FTD. For individuals with PPA, the impairments with communication are rooted in the breakdown of the language system, which can result in significant word-finding problems as well as difficulties understanding single words, simple commands, and basic conversations (for a review see Khayum, Wieneke, Rogalski, Robinson, & O'Hara, 2012). For individuals with bvFTD, the communication impairments are initially more related to the blunted expression and comprehension of emotional information [e.g., prosody, empathy] resulting in problems with the pragmatics of communication. As the disease progresses, individuals with bvFTD may begin to develop expressive language impairments along with impairments in the initiation of communication in any form (i.e., verbal or gestural). Speech-Language Pathologists (SLP) have expertise in analyzing the break-downs in communication to determine how best to compensate for the particular communication problem through word-finding strategies and partner-supported communication techniques.

The main goal of SLP services is to develop strategies to help the person with FTD maximize his/her communication skills, not to regain lost language ability. This is in contrast to aphasia therapy services used in cerebrovascular disease, for example, where it may be possible to re-gain lost language function. Speech-Language Pathologists should tailor the communication skills to the individual needs of the person with FTD and the social environment within which he/she lives, as well as to the stage of the disease. Interventions can include development of skills for facilitating communication of information, including basic wants and needs as well as complex concepts. It also can address some of the impairments in pragmatics that occur particularly with bvFTD which interfere with interpersonal relationships. Assisting the individual and family and/or caregiver with skills for successful interactions will promote positive social relationships and assist with daily life task completion. In anticipation of further decline in communication skills, SLP services may shift to teaching the use of communication tools, such as augmentative communication devices or communication boards (for a review see Fried-Oken, Beukelman, & Hux, 2012; Khayum, et al., 2012). Overall, SLP interventions are focused on developing effective communication skills and strategies proactively, which allows the individual with FTD and his/her caregiver to become proficient in the use of strategies and augmentative devices prior to when they need to be relied upon. The plan for treatment should be a fluid process so that it can adapt along with the emergence of symptoms over the course of disease.

Anecdotal reports indicate that SLP services are useful for individuals with FTD, especially if therapy is maintained throughout the course of disease and realistic goals are set that are patient-centered and include the family and/or caregiver (see Khayum, et al., 2012 for a review for PPA). However, recent evidence suggest that persons with FTD are not being referred for and thus not receiving SLP services as readily as persons with other neurological diseases (e.g. stroke), despite the primary language impairment that is present in PPA (Taylor, Kingma, Croot, & Nickels, 2009). This is likely due to the misinformation that Speech-Language Pathology services are not appropriate because of the progressive nature of the disease. Continued efforts are needed to dispel this notion, to improve awareness about the relevance of language therapy for FTD patients, and to guide SLP's on effective strategies for providing therapy in neurodegenerative cases.

Self-care impairments

Completion of self-care tasks, such as dressing, bathing, and feeding, decline over the course of FTD quite typically secondary to the declines in cognitive functioning, such as impairments in initiation and perseverance, impulsivity, and perseveration (Kipps, Mioshi, & Hodges, 2009). Although the individual may be able to state what step needs to be taken

or that he/she needs to perform a task, the individual may have difficulty in starting the sequence of actions to complete a task, persisting with the action once initiated, starting a task without thinking through the steps first, or perseverating on a particular step. Occupational Therapy (OT) is a rehabilitation field that focuses on assisting individuals in tailoring the self-care tasks to maximize successful completion with the lowest level of assistance as possible. Occupational Therapists have unique training in how to break down activities of daily life, not just self-care tasks, to identify the constituent steps of task completion in order to assist an individual in learning to complete the task by compensating for the impairments. Through participation in OT services, persons with FTD and their caregivers would learn skills and strategies to approach the activities of daily life, thus reducing the burden of care as well as facilitating that the individual with FTD stays engaged in life activities as long as possible. As the disease progresses, OT interventions can be tailored to address the functional impairments as they develop (for a review and clinical practice guidelines see Padilla, 2011).

Behavioral control and social competency impairments

Degeneration of the frontal systems results in behavioral control and monitoring impairments that can dramatically impact the individual's ability to appropriately interact, initiation, and follow-through in daily life. As such, individuals with the behavioral variant of FTD are at risk for both life management problems as well as difficulty maintaining interpersonal relationships. These individuals tend to make disinhibited comments that are off-color or are offensive, may engage in seemingly uncontrolled eating behavior or persist in ritualized behaviors, or may not readily engage in behavior or express emotions, leaving others to think they are uninterested in social interactions and participation in life activities. These are some of the most stressful symptoms for the family and care-givers, whereas the person with bvFTD is typically unaware that there is anything aberrant about his/her behavior, which only adds to the frustration of others. Although research continues to explore pharmacologic options for addressing these behavioral issues, there are currently extremely limited options (for a review, see O'Brien & Burns, 2011). Behavioral modification interventions are alternatives offered by Psychologists, Occupational Therapists, and Geriatric Nurses, among others to assist caregivers in learning strategies to positively impact behavioral control. The Antecedent-Behavior-Consequence Model has been applied to teach caregivers how to respond and modify the behavioral response of the person with FTD over time (for an example see Merrilees, 2007). Another approach to behavioral modification is focused on meeting the unmet needs of the individual as the unmet needs are viewed as triggers for some behavioral issues. Some research supports that interventions based upon this approach is effective in reducing behavioral problems, such as agitation and aggression in other dementia populations (Davison, Hudgson, McCabe, George, & Buchanan, 2007), which also may be an option for individuals with bvFTD. Advancing Caregiver Training (ACT) adds additional components to the "unmet needs" approach by also focusing caregiver training on the role that their own status (stress level, communication style) and the status of the environment (e.g., presence of clutter or hazards) can play as triggers for behavioral problems (Gitlin, Winter, Dennis, Hodgson, & Hauck, 2010). ACT has been found to result in a reduction in behavior problems in the individual with dementia and improved confidence level and reduced sense of burden for caregivers (Gitlin, et al., 2010). Although research needs to continue to investigate the effectiveness of behavioral interventions to assist in reducing the behavioral issues seen in individuals with FTD, it is clear that caregivers benefit from the guidance on how to interpret the behavior from their love-ones with FTD and ways that they can potentially positively impact the daily life functioning. Thus, referral to clinicians with expertise in behavioral modification would be of benefit for individuals with FTD and their caregivers.

Environmental Modifications for Facilitating Life Participation

Human behavior is guided in many ways by the cues and structure that is present in the daily life environment at home, at work, and out in the community. However, individuals with bvFTD have difficulty processing more subtle environmental cues, including the facial expressions and emotional tone of others leading them to have difficulty behaving appropriately (Lough & Hodges, 2002). Matching to a person's areas of competencies and the demands of the environment can assist the individual in functioning at a higher level. Analysis of the qualities of the environment, such as familiarity, physical attributes, and level of structure can assist in identifying environmental factors that may facilitate or hinder an individual's functioning depending upon his/her abilities. Rehabilitation clinicians, particularly Occupational Therapists, but also Speech-Language Pathologists and Rehabilitation Psychologists have the expertise for generating ideas to structure the daily life schedule and living environment to promote self-reliance and safety for individuals with dementia (Gitlin, Hauck, Dennis, & Winter, 2005). Interventions can be focused on educating and training caregivers to modify the daily life structure and physical environment to reduce safety risks and compensate for cognitive impairments to improve daily life functioning.

Modifying and structuring the living environment can reduce some of the safety concerns associated with bvFTD, such as wandering, perseverative behaviors, and falls (for a review see Day, Carreon, & Stump, 2000). Use of alarms on beds and doors can assist caregivers to monitor if a person with FTD has gotten up from bed or is attempting to leave the house. Signage can alert a person who has bvFTD to avoid a dangerous area or item. Furniture can be arranged to facilitate the flow of movement away from stairs or to reduce the risk of falling. Rehabilitation professionals can assist in identifying the possible interplay between the individual's physical and cognitive impairments and the environmental demands and make recommendations for modifying the environment to increase the safety of the environment (for a review see Buchanan, et al., 2011).

Structuring the environment to facilitate participation in activities, or understanding social cues, for remembering tasks to do, and for following through with self-care activities is a key part of assisting the person with FTD live an active, engaged life with the lowest level of assistance possible. Helping caregivers to set up a set schedule for bed-times and rise-times and meals times and other consistent daily activities can assist the person with bvFTD to learn the schedule over time and reduce the chance for agitation secondary to physiological triggers, such as hunger or sleepiness. The physical environment can be structured to reduce the interaction of the environmental characteristics with the cognitive impairments and promote task completion. For example, if a person has difficulty resisting distraction secondary to attention problems, then structuring the environment to reduce background noise and visual distractions can facilitate that the person can successfully stay focused on a task and thus complete the task. Avoiding crowded, noisy environments may allow for better opportunity for communication and avoid frustration for individuals with PPA. For example, when going out to eat the family may ask to be seated near the edge rather than the middle of the dining area. Similarly, reminding friends and family to speak slowly and avoid group conversations at social gatherings may also be helpful. Over time and with progression of the disease, the salience of natural environmental cues declines leading the individual with FTD, especially those with bvFTD, to behave in ways that does not match the social or environmental context. Rehabilitation clinicians can assist caregivers in learning to provide appropriate prompts and assistance and set up environmental cues and structure to assist the individual in behaving in ways that align with the social context. Training caregivers to identify and rectify unmet needs of individual with FTD, manage

their own stress, and reduce environmental barriers results in fewer behavioral problems in the person with bvFTD and improves life functioning (Gitlin, et al., 2010).

Evidence of Language Therapy Intervention Effects for Maintenance of Brain Functioning

In addition to being a relatively rare disorder, the neurodegenerative nature of the disease, heterogeneity of clinical symptoms, and variable rate of disease progression are among the many challenges associated with planning therapeutic interventions in PPA. As a result, most intervention studies have been case reports or small cohorts providing proof of concept that a particular therapeutic approach may be useful (for a review see Croot, Nickels, Laurence, & Manning, 2009). Therapies focused on improving disintegrating language function have primarily focused on strategies to improve word retrieval. A review on treatment of anomia in semantic dementia suggests there is potential for new lexical learning and to slow the progression of anomia over time (Henry, Beeson, & Rapcsak, 2008). Not surprisingly, the authors conclude that the greatest learning potential is available at the early stages of disease before substantial disintegration of the language network has occurred. Novel speech therapy treatment strategies are emerging for specific symptoms commonly associated with PPA. For example, a case study report suggests that multisyllabic word production deficits, a symptom most commonly seen in the agrammatic variant of PPA, may be improved by using a treatment regimen requiring structured oral reading of text and may have effects that are lasting and generalizable to other language deficits (Henry et al., 2012). Preliminary evidence from patients with the semantic variant of PPA suggests that errorless learning strategies may be more beneficial than errorful learning strategies (Jokel & Anderson, 2012). An intensive 2-week semantically based intervention was successful at producing lasting and generalizable effects of improved lexical retrieval over a 6-month period in a patient with the logopenic variant of PPA (Beeson et al., 2011). Though the type of therapy and patients targeted differ by study, outcomes commonly demonstrate learning is possible; however, the generalizability and maintenance of learning is variable.

As more systematic studies of aphasia therapy develop, it will be important that researchers clearly specify the goal of the intervention, the ideal length of therapy, the projected duration of benefit, the generalizability of the treatment to other aspects of language and cognition, and the disease-stage at which therapy may be useful. This will assist clinicians in matching the parameters of the intervention to the needs of the individual presenting for treatment. For therapies that do not directly modify the neurobiology of the underlying disease process, it may be unrealistic to expect vast improvement; however, slowing the rate of decline should be counted as a success.

Integration of Caregivers and Families in Rehabilitation Process

With a typical age of onset between 45 and 65 years, the majority of individuals who have FTD are still actively involved in many instrumental life roles, such as parenting and working. The onset of the cognitive, behavioral, motor, and sensory impairments leads the individual with FTD to assume more of a dependent role within the family, changing the family dynamic. Quite frequently, the caregiver is a family member, either a spouse or an adult child who needs to adapt and adjust to the changing roles as well as provide care for an individual with FTD who is behaving differently and has reduced abilities. Recent research suggests that the caregiver burden is greater in FTD than in other progressive dementia because of the communication and behavioral control impairments that are inherent in FTD, but not necessarily part of the symptom picture for other progressive dementias (Mioshi, Bristow, Cook, & Hodges, 2009; Riedijk et al., 2006). Integration of the family and caregiver(s) into the rehabilitation process provides them opportunities for learning about FTD and its impact on life functioning, to put into context the changes that are occurring cognitively, behaviorally, and physically to the individual with FTD, and to learn ways to

cue, prompt, assist and structure the home environment to maximize life functioning for the entire family (Khayum, et al., 2012; Letts et al., 2011). Additionally, caregivers can play a critical role in the rehabilitation process by tracking and reporting symptoms over time so that the interventions can be tailored to needs of the individual with FTD (Jicha, 2011). Finally, there is evidence that interventions can help to reduce caregiver distress by providing these individuals with information, guidance, and training to best support the individual with FTD (e.g., Letts, et al., 2011).

The goal for many families is to ensure that the individual with FTD be able to live at home and engage in typical daily life activities for as long as possible. Accomplishment of that goal requires that the family members are well educated by the clinicians about the behavioral features of FTD, guarding against the typical conclusions that the behaviors are intentional or deliberate (Mioshi, et al., 2009). Caregivers must receive training on how to effectively communicate with their loved-one with FTD when the language impairments are part of the symptom picture. They must learn how to appropriately prompt and provide feedback when the individual with FTD does not follow-through with task completion or does not follow social convention when interacting socially. This type of training is a core piece of neurorehabilitation provided by Speech-Language Pathologists, Occupational Therapists, and Rehabilitation Psychologists, for example. A spirit of teamwork can be promoted between the individual with FTD and his/her family and/or care-giver so that they can work together towards the goal of living life in the desired environment for as long as possible without defensiveness, blame, or emotional turmoil degrading the relationship. Integration of the family members and care-givers into the rehabilitation process ensures that everyone has proper context, understanding, and skills for assisting the individual with FTD in engaging appropriately and fully in life as possible.

Community-Based Activities: Role of Support and Activity Groups

Awareness of non-Alzheimer's dementias is increasing in research and also in the community at large through development of support groups, educational conferences and the efforts of nonprofit organizations, such as the Association for Frontotemporal Degeneration (AFTD), the National Aphasia Association (NAA), and the Alzheimer's Association. In fact, over the past year, there were at least four educational conferences for health care professionals and caregivers across the United States at Emory University, Northwestern University, University of North Carolina (UNC), and the University of Pennsylvania. Banks and colleagues (2006) provided practical considerations for organizing and implementing a conference for caregivers and individuals with a diagnosis of PPA and bvFTD, which may be useful for the development of future meetings.

Support groups are an essential part of the care plan, which offer an empathetic environment for family caregivers to share with others who have similar life experiences. Since bvFTD and PPA are less common than the clinical amnesic dementia associated with Alzheimer's disease, the number of support groups is also fewer, making it difficult for individuals to find disease-specific support groups in their area. The Alzheimer's Association and AFTD are willing to help individuals learn how to begin, sustain and maintain a group that specifically offers support to those caring for someone with FTD and PPA. In addition, online support groups and internet-based chat rooms provide alternative methods for caregivers to find support.

If it is not possible find or start a support group specific to PPA/bvFTD, a young-onset dementia support group, such as early-onset Alzheimer's disease, may be a good match. These individuals will be experiencing similar life challenges as a consequence of the disease and so the topics discussed will likely be of interest and helpful for individuals with

PPA/bvFTD and their families (e.g. school-age children at home, financial/legal planning concerns, social security disability, leaving the workplace in the prime of their career).

Activity-based programs (structured meaningful activities) may provide improved quality of life for individuals with FTD. These programs may not necessarily target treatment of language or behavioral symptoms, but instead provide cognitive stimulation and may improve mood and accommodate those who would have difficulty participating in traditional talk-based formats. This alternative model of support was recently piloted at Northwestern University where individuals diagnosed with PPA participated in a group utilizing multiple formats including painting, horticulture class, and improvisation session. Results were positive and suggest individuals with PPA may show a quality of life benefit from participation in activity-based support groups (Rogalski, O'Hara, Wieneke, & Morhardt, 2010; Wieneke, O'Hara, Rogalski, & Morhardt, 2012). Dance or music therapy as well as yoga, meditation and mindfulness programs may also be helpful for this population.

For individuals with bvFTD, traditional talk-based format support groups pose a challenge because disease awareness is often poor and inappropriate social behaviors may interfere with productive conversations. However, activity-based groups, such as those aimed at improving social participation through reminiscence or life-story approached have been shown to have positive outcomes, including improved sense of well-being and life satisfaction (for a review see Letts, et al., 2011).

Summary and Future Directions

Individuals with FTD face unique challenges secondary to the cognitive-behavioral, motor, and sensory decline that can occur with the disease process. Because these individuals are relatively young and engaged in important life roles, such as working and parenting at the time of disease onset, the need for interventions to assist them in maintaining functioning for as long as possible is crucial. There are no known disease-modifying interventions and very limited medical interventions that have been found to assist in the management of the symptoms. Interventions offered by neurorehabilitation specialists and community-based activities offer individuals with bvFTD/PPA interventions and services to improve life functioning and life satisfaction. Educational conferences and support groups provide an opportunity for caregivers and individuals with a diagnosis to meet others facing similar family and life situations so that they can learn from each other and feel less isolated. It is hoped that medical professionals who routinely provide diagnostic services for identifying individuals with bvFTD/PPA will incorporate referrals to neurorehabilitation services and community-based activities as part of routine care. Future research should focus on evaluating the efficacy of these types of behavioral and community-based interventions, including investigating how behavioral interventions help to reduce the burden of care, morbidity, and the reliance on medical care (i.e., health service utilization).

Acknowledgments

The authors alone are responsible for the content and writing of this paper. Dr. Kortte's work is supported in part by the NIH (RO1NS047691-05A2) from the National Institute of Neurological Disorders and Stroke. Dr. Rogalski's work is supported in part by the NIH (DC008552 from the National Institute on Deafness and other Communication Disorders (NIDCD), 1R01NS075075-01A1 from the National Institute of Neurological Disorders and Stroke, AG13854 (Alzheimer Disease Center) from the National Institute on Aging, and 5KL2RR025740 from the National Center for Research Resources, and is now at the National Center for Advancing Translational Sciences, Grant 8KL2TR000107. The content is solely the responsibility of the authors and does not necessarily represent the official views of the NIH.

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Take home points

Behavioral interventions should be a standard part of the care offered to individuals with bvFTD and PPA.

Neurorehabilitation interventions and community-based activities have the potential to benefit both the individual with FTD and the caregiver.

Future Direction

Future research should focus on evaluating the efficacy of these types of behavioral and community-based interventions, including investigating how behavioral interventions help to reduce the burden of care, morbidity, and the reliance on medical care (i.e., health service utilization).

Education for medical professionals about the benefits of non-pharmacologic therapies is needed to improve referral rates to these programs and to help alleviate the burden of care on caregivers and society.