NIH Public Access
Author Manuscript
J Gerontol Nurs. Author manuscript; available in PMC 2013 May 01.

Published in final edited form as:
J Gerontol Nurs. 2013 March ; 39(3): 10–14. doi:10.3928/00989134-20130131-02.

Managing Differences:

Care of the Person with Frontotemporal Degeneration

Geri R. Hall, PhD, ARNP, GCNS, FAAN,
Advanced Practice Nurse, Banner Alzheimer's Institute, Phoenix, Arizona

Jill Shapira, PhD, APRN-BC,
Adult Nurse Practitioner, University of California Los Angeles Neurological Services, Los Angeles, California

Maribeth Gallagher, DNP, PMHNP-BC, and
Director, Dementia Programs, Hospice of the Valley, Phoenix, Arizona

Sharon S. Denny, MA
Program Director, The Association for Frontotemporal Degeneration, Radnor, Pennsylvania

Abstract

Caring for people with non-Alzheimer's dementias is particularly challenging for families and care providers. This is especially true for those with frontotemporal degeneration (FTD) who exhibit profound changes in personality, behavior, language, and movement. Initial symptoms are often misdiagnosed as psychiatric disorders or early-onset Alzheimer's disease, and typically do not respond to pharmacological and nonpharmacological interventions designed for people with other dementias. Using individual examples, this article illustrates common features of two subtypes of FTD: behavioral variant FTD and non-fluent primary progressive aphasia.

Frontotemporal degeneration (FTD) represents a leading cause of early-onset dementia among individuals younger than 65, although onset of symptoms can occur as early as age 21 and as late as 80. It is estimated that FTD affects approximately 50,000 to 60,000 Americans (The Association for Frontotemporal Degeneration, 2012). Life expectancy is 7 to 13 years from disease onset when cases do not involve motor-neuron disease (Onyike, 2011).

The hallmark features of FTD include gradual, progressive decline in personality, behavior, and/or language, with associated changes in motor function among some individuals (Rascovsky et al., 2011). FTD results from progressive damage to the anterior temporal, frontal, or both lobes of the brain due to a spectrum of pathological and genetic disorders. Symptom presentations vary based on the area of the brain affected, although the subtypes of FTD share many clinical characteristics. Functional losses are the result of decreased executive function and may include disinhibited and impulsive behavior, apathy, diminished empathy, and a lack of concern about the disease (Grossman, 2002; Merrilees & Ketelle, 2010; Mendez & Shapira, 2011). Some individuals have significant language impairment. Subsets of individuals develop movement disorders with Parkinsonism such as progressive supranuclear palsy, amyotropic lateral sclerosis, or corticobasal degeneration.

Copyright © SLACK Incorporated

Address correspondence to Geri R. Hall, PhD, ARNP, GCNS, FAAN, Advanced Practice Nurse, Banner Alzheimer's Institute, 901 East Willetta Street, Phoenix, AZ 85006; ghall6872@aol.com.

The authors disclose no conflicts of interest, financial or otherwise.

NIH-PA Author Manuscript
NIH-PA Author Manuscript
NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

TYPES OF FTD

FTD is divided into three types, depending on the earliest presentation of symptoms:

- Progressive behavioral and personality decline—marked by changes in behavior, emotions, concentration, attention, reasoning and judgment, ability to inhibit inappropriate actions, and the capacity for empathy.
- Progressive language decline—characterized by early changes in ability to speak, understand spoken and written language, and write.
- Progressive motor decline—marked by difficulty with movement including motor planning (apraxia), development of tremor, weakness, poor coordination, falls, gait change, and poor coordination (National Institute on Aging, 2010).

DETECTING FTD

Most individuals with FTD perform well on standard cognitive screens and do not suffer from memory loss or visuospatial impairment until the more advanced stage of the disease. Because they score well on most cognitive tests, are often a younger age, and typically have a lack of insight, individuals with FTD are often misdiagnosed as having psychiatric illness, personality disorders, eccentricity, or mild cognitive impairment (Rascovsky et al., 2011). Delays in accurate diagnosis and increasingly impaired judgment contribute to significant problems with issues of capacity, consent, and implementation of advance directives.

Nurses care for people with FTD in a variety of settings including ambulatory clinics, acute psychiatric units, in-home care, adult day programs, and residential long-term care. Individuals with FTD are often young, more physically active, and may exhibit disruptive, angry, and compulsive behaviors. As a general rule, people with FTD do not respond to well-established, evidence-based interventions for Alzheimer's-type dementias (Arvanitakis, 2010), including cholinesterase inhibitors such as donepezil (Aricept®) (Mendez, Shapira, McMurtray, & Licht, 2007) or memantine (Namenda®) (Boxer et al., 2012). In addition, behavioral management strategies used for AD, such as distraction, have limited efficacy for FTD. The purpose of this article is to use individual examples to introduce nurses to concepts of care in two common forms of FTD: behavioral variant FTD (bvFTD) and non-fluent primary progressive aphasia (nfPPA).

INDIVIDUAL EXAMPLE 1: BEHAVIORAL VARIANT FTD

The salient features of bvFTD include profound alterations in personality and behavior causing disruption in social relationships. Intact frontal lobes facilitate both "executive" abilities to design, organize, and perform activities and also help modify emotions and behaviors to fit socially appropriate norms. People with bvFTD exhibit disorganized, apathetic, and disinhibited behaviors, and often develop compulsive thoughts and ritualistic and rigid behaviors.

Mild Disease

Jeff (pseudonym) began to experience difficulty in his work as a certified public accountant at age 48. He made mistakes completing forms, missed appointments with clients, and was accused of sexual harassment after grabbing and kissing a colleague. He divorced his wife of 20 years while she was being treated for breast cancer and moved to a singles apartment without saying farewell to their 15-year-old son. Two years later, Jeff's elderly parents pursued neurological evaluation after they visited him and discovered that his diet consisted solely of potato chips, ice cream, and candy bars; he had subscriptions to 24 magazines arranged in disorganized piles; and his dog was dehydrated from lack of water.

NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

Diagnosed with bvFTD, a family meeting with the nurse practitioner was scheduled. Jeff complacently agreed to live with his parents although he insisted nothing was wrong. Jeff's family joined support groups. Jeff agreed to wear a Medic-Alert®+Safe Return® identification bracelet (http://www.alz.org/care/dementia-medic-alert-safe-return.asp) and had a global positioning satellite (GPS) device placed in his shoe after he disappeared for several hours. The treatment team assisted Jeff's family to obtain retroactive disability benefits, as his inappropriate behavior was due to a neurological disease. His living situation improved with the monitoring and supervision provided by his parents. Although he had no independent interests or hobbies other than taking long walks daily, he quietly accompanied his parents to concerts, lectures, and dinners with their friends.

Moderate Disease

Over the next 18 months, Jeff's disease progressed. He needed to be led into the shower and would shave only when the electric razor was placed in his hand. Jeff occasionally experienced urinary and bowel incontinence, particularly when in public. Urinary tract infection and constipation were ruled out and Jeff's parents decided to use protective garments for his incontinence. He continued to take long walks, but due to compulsive walking he was unable to stop and return on his own. On two occasions the family solicited assistance from the police to find and return him. Several neighbors complained that he took newspapers, children's toys, and mail from their properties. During the clinic evaluation, Jeff sat quietly and answered questions with either "yes" or "no" or by shaking his head. The family was counseled to have 24/7 supervision and activities for him, including participation in an adult day program. The family made sure he had the charged GPS tracker on him at all times.

Advanced Disease

Jeff was placed in a residential care facility 6 months later, as his parents were physically unable to bathe and change him. Although his parents were understandably concerned about the move, Jeff had no difficulty adjusting to the new environment. He continued to comply with directions and allowed staff to help him bathe and dress. Several weeks after admission, Jeff tried to force several female staff members onto his bed, but did not resist when firmly told "no."

He no longer spoke, but would occasionally shake or nod his head. During this time, Jeff started picking his nails until they bled and grabbing food from the plates of other residents. His physician prescribed a selective serotonin reuptake inhibitor (SSRI) to help manage these repetitive behaviors, and a staff member was assigned to monitor him at mealtimes. Staff also explored activities that would engage Jeff and try to distract him from picking his nails. They found he would stop when given picture and letter cards to sort. He was more likely to participate when activities were individualized and a staff member sat beside him to get him started.

Throughout the course of his disease, Jeff exhibited many of the most common characteristics of bvFTD (Miller, 2012). He lacked insight about his disability. He had trouble organizing and completing tasks (executive impairment). He made sexual advances to a colleague and took neighbors' mail and newspapers (disinhibited behavior/poor impulse control). He ordered 24 different magazine subscriptions, grabbed food from others, and repetitively picked at his nails (compulsive and repetitive behavior). Jeff divorced his wife when ill, left his son without farewell, and ignored his dog's thirst (lack of empathy). As his disease progressed, Jeff developed incontinence and lost the ability to speak.

NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

INDIVIDUAL EXAMPLE 2: NON-FLUENT PRIMARY PROGRESSIVE APHASIA

In nfPPA, a gradual progressive impairment of language production, object naming, and syntax occurs. Individuals lose the ability to use spoken language because of an oral apraxia (planning and sequencing complex voluntary activity). Initially, they know what they want to say but are unable to sequence movements to create speech. This progresses to include additional receptive language impairment and cognitive and behavioral symptoms.

Mild Disease

Grace (pseudonym) was a 57-year-old single nurse diagnosed with nfPPA after she began to speak more slowly with halting speech and became frustrated when trying to give report or work with her clients. She demonstrated many of the typical signs of mild disease (Miller, 2012). Although she knew what she wanted to say, she was unable to coordinate the muscles to produce the sound. Because she maintained the ability to understand verbal and written language, she was able to communicate using her computer, but became increasingly quiet and was forced to apply for disability. Grace was aware of her condition and demonstrated extreme anxiety and frustration when trying to communicate with family and friends.

After her diagnosis, Grace was referred to an advanced practice nurse (APN) to coordinate her care and a speech pathologist who helped her to find adaptive devices. She continued to live alone and drive, wearing a Medic-Alert®+Safe Return® bracelet. She carried a letter from her provider explaining her language deficit in case she encountered situations where she was unable to explain herself. The speech pathologist located computerized applications to assist with communication for her pad-type device, and measures were undertaken with police and fire departments to respond in case of emergency. No medications were prescribed for Grace at that visit.

Moderate Disease

Over the next year, Grace had increased problems with producing speech, compensated with her adaptive devices and using “yes/no” responses. Her reading diminished but she could use her adaptive pad as a language board. Her care team noted that Grace was beginning to experience receptive language difficulties. No longer able to answer the phone, Grace could not call for help when a solicitor forced his way into her house. Her sister convinced Grace to move in with her but the relocation caused confusion, depression, and insecurity about being left alone during the day. The APN referred Grace to an adult day program to provide structure while her sister was at work. The APN ordered occupational therapy every 6 months to evaluate function and design leisure activities. Grace was able to use a picture board and enjoyed jigsaw and find-a-word puzzles. Physical therapy was ordered to increase postural stability and control as Grace began to develop motor symptoms and rigidity.

Over time, Grace’s behavior was becoming disinhibited. She had spontaneous bowel movements in the supermarket and refused to allow clean-up. In restaurants, she began to stuff her mouth with food and spit it out. At night she raided the pantry and refrigerator. Fearful she might choke in the night, the APN recommended locks for food storage areas. A baseline swallowing study was ordered with normal results.

Advanced Disease

As Grace’s behavioral symptoms worsened, her sister placed Grace in a memory care unit of a nursing facility. Grace also developed pica, compulsive eating of non-food items. Found eating tea bags, napkins, toiletries, and soap, staff increased monitoring of her actions, never leaving items in sight that could be mistakenly eaten. She continued to stuff her mouth when

NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

eating and began to choke. Follow-up swallowing studies indicated the need for a soft consistency diet, and staff had to carefully supervise all eating and medication administration. The staff tried to keep Grace engaged in an array of activities that brought her pleasure and distracted her from risky behaviors.

Grace developed gait apraxia and required a wheelchair when she was out of bed. Loss of ambulation resulted in her screaming for up to 24 hours per day. Numerous interventions and medications, including antipsychotic, SSRI-type antidepressant, mood-stabilizing (valproic acid) and anxiolytic agents, and memantine were tried with limited success.

The APN recognized that Grace demonstrated many of the characteristics of nfPPA that occur near the end of life. She was bed bound, completely dependent on others for all activities of daily living, incontinent of bowel/bladder, spoke only a few words, demonstrated muscle wasting, choking, and coughing on food/liquid/saliva, and had recurrent infections (Miller, 2012). The APN met with Grace's sister to discuss Grace's significant decline and educated her on the services offered by hospice. They agreed that Grace would choose comfort as the primary goal of care and decided to initiate a referral to hospice.

End-of-Life/Hospice Care

Medicare's hospice eligibility criteria for dementia pose challenges to timely services for people with FTD because the criteria were built on the progression of AD. Although individuals with FTD exhibit different symptoms from each other in the earlier stages depending on the variant, all forms of FTD tend to appear similar at end of life.

A hospice nurse visited Grace and her sister. Reviewing her medical records, the nurse determined that Grace fulfilled Medicare's criteria for hospice eligibility (National Hospice Organization, 1996). The nurse explored the goals of care with Grace's sister. Although she never had a discussion with Grace about specific details regarding health care decisions, her sister used her best substituted judgment to identify that Grace would not choose aggressive interventions to prolong her life when there was perhaps little benefit to improve her condition. Each item on the health care directives was discussed with the support of the hospice staff so that Grace's sister could make informed decisions to honor Grace's wishes. The completed document clearly communicated that Grace did not choose resuscitation, hospitalization, antibiotic agents, or a feeding tube. Instead, the goals focused on aggressive comfort care so that Grace could live out her days safely, while comfortable in the company of those who knew her best.

Grace's medication regimen was simplified to include only essential medications to maximize comfort. Routine acetaminophen (e.g., Tylenol®) was prescribed for stiffness and chronic pain issues. The staff used a non-verbal pain assessment tool to evaluate comfort.

The interdisciplinary hospice team focused on optimizing Grace's quality of life by providing daily sensory pleasures based on those she enjoyed throughout her lifetime. Favorite music, prayers, and poetry kept her engaged for periods of time throughout the day. Grace's diet was liberalized and included small frequent offerings of items she enjoyed most and tolerated safely. Her sister was encouraged to bring in familiar items from the past such as photographs and other touchstones to reminisce meaningful moments of her life.

Staff reported that Grace's screaming episodes, although not completely eradicated, diminished significantly within days after the aforementioned pharmacological and nonpharmacological comfort measures were initiated. Grace developed aspiration pneumonia. She was not admitted to the hospital or administered antibiotic agents. Her

NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

comfort was maximized using medications to address fever, pain, and restlessness, while her favorite hymns softly played in the background to create a serene and supportive environment for everyone in the room. Shortly thereafter, Grace died a peaceful death surrounded by her sister, some favorite long-term facility staff, and her hospice nurse. The hospice team supported Grace's sister and the nursing facility staff through the entire process and referred the sister to the bereavement specialist for 13 months of follow-up care.

IMPLICATIONS

Wherever they practice, nurses presented with a person who falls outside the expected behavior for AD-type dementia should consider the person may have FTD. The majority of interventions in FTD are nonpharmacological nursing measures, including continuous assessment, monitoring the person's response to stimulus, structuring the routine, and providing for safety in a least restrictive manner. In every setting, nurses evaluate the need for other disciplines such as physical therapy, activities, or speech pathology and work to assure follow-through on interdisciplinary recommendations. Most important, nurses assess and intervene using specific activities and behavioral techniques that minimize the potential for conflict and safety hazards while maximizing remaining cognitive strengths.

Resources for FTD Information

- "Frontotemporal Disorders: Information for Patients, Families, and Caregivers," a free educational booklet for parents and families from the National Institute on Aging and National Institutes of Health at http://www.nia.nih.gov/sites/default/files/frontotemporal_disorders_information_for_patients_families_and_caregivers_0.pdf
- Education and resources for families and professionals are available through The Association for Frontotemporal Dementia at http://www.theaftd.org.

CONCLUSION

Prominent symptoms associated with FTD include inappropriate social and emotional behavior and impaired communication in individuals who are younger and more physically active than people with AD. These features impact both the usual ways nurses relate to people in their care and the efficacy of interventions established for more typical Alzheimer's-type dementias. Understanding the anatomical underpinnings of these clinical presentations allows nurses to target disease-specific behaviors while maximizing remaining cognitive strengths.

Due to relatively small numbers of people with FTD, there are currently no published evidence-based care practices. Using information developed by nurses and families for The Association for Frontotemporal Degeneration and FTD researchers can help nurses and family members work together to create targeted strategies of behavior management and provide family support. Prompt recognition of the advanced stages of FTD permits timely referral to hospice services.

Acknowledgments

Dr. Shapira is supported by National Institutes of Health grant R01AG03499 (PI: M.F. Mendez).

REFERENCES

Arvanitakis Z. Update on frontotemporal dementia. Neurologist. 2010; 16:16–22. [PubMed: 20065792]

NIH-PA Author Manuscript

NIH-PA Author Manuscript

NIH-PA Author Manuscript

The Association for Frontotemporal Degeneration. Frontotemporal degeneration. 2012. Retrieved from http://www.theaftd.org/frontotemporal-degeneration/ftd-overview

Boxer, A.; Knopman, D.; Kaufer, D.; Grossman, M.; Onyike, C.; Graf-Radford, N.; Miller, B. A 26-week, randomized, multicenter, placebo controlled trial of memantine for behavioral variant frontotemporal dementia and semantic variant primary progressive aphasia; Manchester, England. Paper presented at the Eighth International Conference on Frontotemporal Dementias; 2012 Sep.

Grossman M. Frontotemporal dementia: An update. Journal of International Neuropsychological Society. 2002; 8:566–583.

Mendez MF, Shapira JS. Loss of emotional insight in behavioral variant frontotemporal dementia or "frontal anosodiaphoria". Consciousness and Cognition. 2011; 20:1690–1696. [PubMed: 21959203]

Mendez MF, Shapira JS, McMurtray A, Licht E. Preliminary findings: Behavioral worsening on donepezil in patients with frontotemporal dementia. American Journal of Geriatric Psychiatry. 2007; 15:84–87. [PubMed: 17194818]

Merrilees J, Ketelle R. Advanced practice nursing: Meeting the caregiving challenges for families of persons with frontotemporal dementia. Clinical Nurse Specialist. 2010; 24:245–251. [PubMed: 20716977]

Miller, B. Frontotemporal dementia: Disease progression. 2012. Retrieved from http://memory.ucsf.edu/ftd/overview/ftd/progression/multiple

National Hospice Organization. Medical guidelines for determining prognosis in selected non-cancer diseases. 2nd ed. Arlington, VA: Author; 1996.

National Institute on Aging. Fronto-temporal disorders: Information for patients, families, and caregivers. 2010. Retrieved from http://www.nia.nih.gov/sites/default/files/frontotemporal_disorders_information_for_patients_families_and_caregivers_0.pdf

Onyike CU. What is the life expectancy in frontotemporal lobar degeneration? Neuroepidemiology. 2011; 37:166–167. [PubMed: 22056969]

Rascovsky K, Hodges JR, Knopman D, Mendez MF, Kramer JH, Neuhaus J, Miller BL. Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia. Brain. 2011; 134:2456–2477. [PubMed: 21810890]