

Long-Term Care of Patients With Frontotemporal Dementia

Jennifer J. Merrilees, RN, MS, and Bruce L. Miller, MD

Frontotemporal dementia (FTD) results from the progressive dysfunction of the frontal and/or temporal lobes of the brain. It is a presenile dementia with a mean age of onset of 52.8 to 56 years.¹ FTD affects men and women equally. The course of FTD has a mean duration of 8 years.² There are no known risk factors for FTD, although approximately 40% of patients have a family history of dementia.³ With this disorder, there are marked changes in personality and behavior. As a result of the profound impact on patients' function, most patients with FTD will eventually require placement in a long-term care setting.

Consensus criteria from 1998 outlines 3 clinical presentations: frontotemporal dementia (FTD), semantic dementia (SD), and progressive nonfluent aphasia (PNFA).⁴ The term frontotemporal lobar degeneration (FTLD) is often used in reference to the 3 syndromes. The clinical features of the 3 subtypes are determined by the region of the brain affected.

Key features of patients with right frontal and bifrontal lobe involvement are behavioral symptoms and personality changes. Patients can exhibit disinhibition and impulsivity. Stereotypic behaviors such as hoarding, impulse buying, and compulsions are also common. Apathy and lack of drive also occur. Personality changes include emotional blunting and blunting of empathy; it is common to confuse these symptoms with depression, midlife crisis, and marital problems. Cognitively, patients with FTD have difficulties with organization, planning, and reasoning. They tend to have limited insight about their behavioral and personality alterations.

Remarkably, sometimes the changes can be viewed as positive. Early disinhibition in one patient resulted in increased assertiveness and self-confidence. She demonstrated a new interest in the stock market and made successful transactions over a period of months. She exhibited a new flare in her style and dress. Her family found her to be more affectionate. As her disease progressed, she exhibited poor judgment, socially inappropriate behaviors, and profound apathy requiring 24-hour supervision and eventual long-term placement.

Right temporal patients also present with behavioral symptoms that can appear strange and psychiatric in nature. They can exhibit disinhibition, irritability, and loss of empathy. Their thinking becomes rigid, stereotyped, and they can dem-

onstrate alterations in dress and hygiene. Hyperorality and ritualistic behaviors around eating can occur. Socially inappropriate behaviors, including theft, offensive speech, assault, and public urination or masturbation, occurred in almost half of patients with FTD with atrophy of the frontal and temporal lobes.⁵

Language dysfunction dominates left-sided involvement, although behavioral symptoms do emerge. Progressive nonfluent aphasia (PNFA) occurs in patients with left frontal involvement and is characterized by diminished ability to express language. Patients experience difficulty with speech initiation, pronunciation, and word finding. Associated clinical features include depression, social withdrawal, and preservation of social graces.¹ Behavioral symptoms do not usually appear until later in the disease.⁶ Semantic dementia is caused by involvement of the left temporal lobe; patients demonstrate progressive loss of knowledge about words and objects. Speech might be fluent, but is characterized by semantic paraphrasias and substitutive phrases. New artistic talents and skills have been observed in some patients.⁷

Patients with FTD typically present with 1 of the 3 different clinical syndromes. Onset of symptoms is insidious and slowly progressive. As the disease progresses, patients can exhibit features of the other clinical syndromes.

Motor abnormalities are common in FTD as a result of the association with 3 other neurodegenerative diseases: corticobasal ganglionic degeneration (CBD), progressive supranuclear palsy (PSP), and motor neuron disease (MND). Conversely, patients with PSP and CBD often develop behavioral symptoms. Apathy and depression have been observed in patients with PSP. Patients with CBD can exhibit personality changes, disinhibition, and irritability.

There is currently no treatment for slowing or reversing FTD. Acetylcholinesterase inhibitors, used to slow progression in Alzheimer's disease, are avoided in FTD for several reasons. In clinical experience, they tend to offer no improvement in cognition and can promote agitation and irritability. Management of FTD is aimed at reduction of problematic behavioral symptoms, health maintenance and prevention of future problems, and adaptation to changes in functional abilities. With disease progression, patients become more functionally dependent. They demonstrate a reduction in speech, eventually becoming mute. Motor abilities decline and swallowing difficulties can occur. Some behavioral symptoms diminish and others surface. Death typically results from factors associated with immobility.

Patients with FTD and their families face a myriad of overarching concerns. FTD causes profound disruption for families as they deal with behavioral, personality, and func-

Department of Neurology, University of California at San Francisco, San Francisco, California

Address correspondence to: Jennifer Merrilees, RN, MS, UCSF Memory and Aging Center, 350 Parnassus Avenue, Suite 706, San Francisco, CA 94143-1207. E-mail: jmerrilees@memory.ucsf.edu

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Table 1. Medications for Neurobehavioral Symptoms of FTD

Drug Category	Suggested Use	Medication Name
Selective serotonin reuptake inhibitors	Impulsivity compulsivity, depression, carbohydrate craving, aggression, anxiety, delusions, irritability	Sertraline, citalopram, fluoxetine, paroxetine
Atypical antipsychotics	Aggression, agitation, hallucinations	Olanzapine, quetiapine, risperidone
Anticonvulsant	Agitation, aggression	Valproic acid
Benzodiazepines	Short term only, eg, before a procedure	Lorazepam
Newer antidepressants	Depression	Venlafaxine, bupropion

FTD = frontotemporal dementia.

tional alterations. One family member described herself as being “crushed by the demands” she faced as the primary caregiver for her mother with FTD.

MANAGEMENT OF BEHAVIORAL SYMPTOMS

Methods to ameliorate behavioral symptoms ideally incorporate strategies that focus on environmental and behavioral manipulation, as well as appropriate use of medication. Although there have been some studies evaluating efficacy of medications for behavioral symptoms in FTD, there have been no systematic evaluations of appropriate environmental and behavioral interventions. It is tempting yet speculative to use interventions for Alzheimer’s disease (AD). Yet, there are important cognitive differences between AD and FTD. Generally, memory is preserved until later in the disease. This memory preservation can be viewed as an asset and should influence future research into behavioral interventions. The fields of psychiatry and neurorehabilitation hold promise as models for research into effective interventions for behavioral symptoms in FTD.

Serotonin deficits are thought to be a cause for behavioral symptoms in FTD; therefore, selective serotonin reuptake inhibitors (SSRIs) are used for many of the behaviors. SSRIs have been shown to be helpful in treating impulsivity, compulsions, depression, and carbohydrate craving.⁸ SSRIs can be helpful with aggression, delusions, and anxiety. Atypical antipsychotics have been used for aggressive behaviors. Valproic acid has use for agitation and aggression. Antipsychotics such as haloperidol are not used in FTD as a result of undesirable side effects. Benzodiazepines are not recommended because of their negative cognitive effects; however, they can be useful as short-term treatment, in selected patients, to complete a necessary test such as magnetic resonance imaging (see Table 1).

HEALTH MAINTENANCE AND PREVENTATIVE CARE

The decline in speech and language ability creates communication challenges for patients with FTD and their families. Clinical experience suggests that speech therapy offers benefits for some patients, yet it is unclear the role that therapy plays in this disease. In particular, the patients with nonfluent progressive aphasia tend to benefit.

Exercise is recommended for all patients. As motor symptoms emerge, patients will be at risk for complications of immobility, falls, and fractures. Rehabilitation principles

aimed at reduction of risk can be used.⁹ Swallowing problems can develop, requiring referral for swallowing assessment. Dietary changes such as substituting thick for thin liquids are often made.

Early planning for incapacity is important in making decisions about future healthcare needs and protection of assets. Clarification of wishes around topics such as resuscitation, life support, and autopsy are ideally made when the patient is able to articulate their wishes.

FUNCTIONAL CHANGES

The patterns of change in the patient’s functional abilities are unique and variable in FTD. Deficits in organizational and planning abilities, poor judgment, and apathy affect job performance, driving, chores, and hobbies. Counseling regarding early retirement, disability, and driving is often necessary. Language difficulties affect job performance as well, although in the absence of executive dysfunction, many of our language patients continue to work successfully. Patients elect to stop working as their language and speech deficits advance. Neglect for personal care and safety risks occur in patients with disabling compulsive behaviors. An obsession with rock collecting made one patient unable to care for her young child. Motor complications cause functional deficits in self-care and increased risk for injury, especially when behavioral symptoms such as impulsivity and stubbornness are present.

DETERMINING THE NEED FOR CARE

Each patient has a unique expression of the disease. Clinical experience shows that factors such as home setting, rural versus urban setting, available resources, quality of family caregiving, and other demands on the family unit all factor into the planning of long-term care needs. Like with all forms of dementia, it is the unique interplay of behavioral symptoms, self-care abilities and needs that often determine whether additional help is needed.

THE TRANSITION TO A NEW LEVEL OF CARE

It is vital to match the needs of the patient with resources of the facility. Families are important sources of information for insight about the patient’s cognitive, behavioral, and physical attributes. Patients with a history of wandering and elopement will need careful attention to safety needs. Strategies currently exist for such patients, including warning alarms on exits, secured grounds, or one-on-one companionship/super-

vision. Restraint use is associated with profound detrimental effects, and their use is not encouraged.^{10,11} Speech and language deficits result in alternative forms of communication. Strict attention to body language and behavioral cues are necessary. It is important to evaluate the patient's activity level and needs to ensure they can be met by the facility.

No standard way of structuring the transition to a different level of care has been studied in this population of patients. Some families have been encouraged to stay away for a period of time to allow for adaptation and acceptance; others spend a lot of time at the new facility. It might be helpful to conduct shorter respite visits as a way of introducing the patient to the new facility. It has been fairly typical for patients to express anger toward the family caregiver for the move to long-term care.

The transition to a long-term care facility is marked by disruption of a familiar routine, new names and faces, and unfamiliar residents. Catastrophic reactions characterized by increasing disorganization, irritability, and agitation occur when the patient becomes overwhelmed and overloaded by demands placed by the environment. Every attempt should be made to decrease the stress for the patient and find acceptable methods for stress reduction. In a few rare cases, a temporary move to a secure unit for medication management has been necessary.

In summary, patients with FTD are characterized by profound changes in their behavior, language, and personality. The course of FTD is heterogeneous and slowly progressive. Patients require assistance and supervision for a myriad of reasons, and most eventually need placement in a long-term

care facility. Caring for this group of patients is nearly always extremely challenging, yet often rewarding. New therapies for FTD are likely to emerge in the coming years, and with these therapies, there will be new, exciting clinical challenges.

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