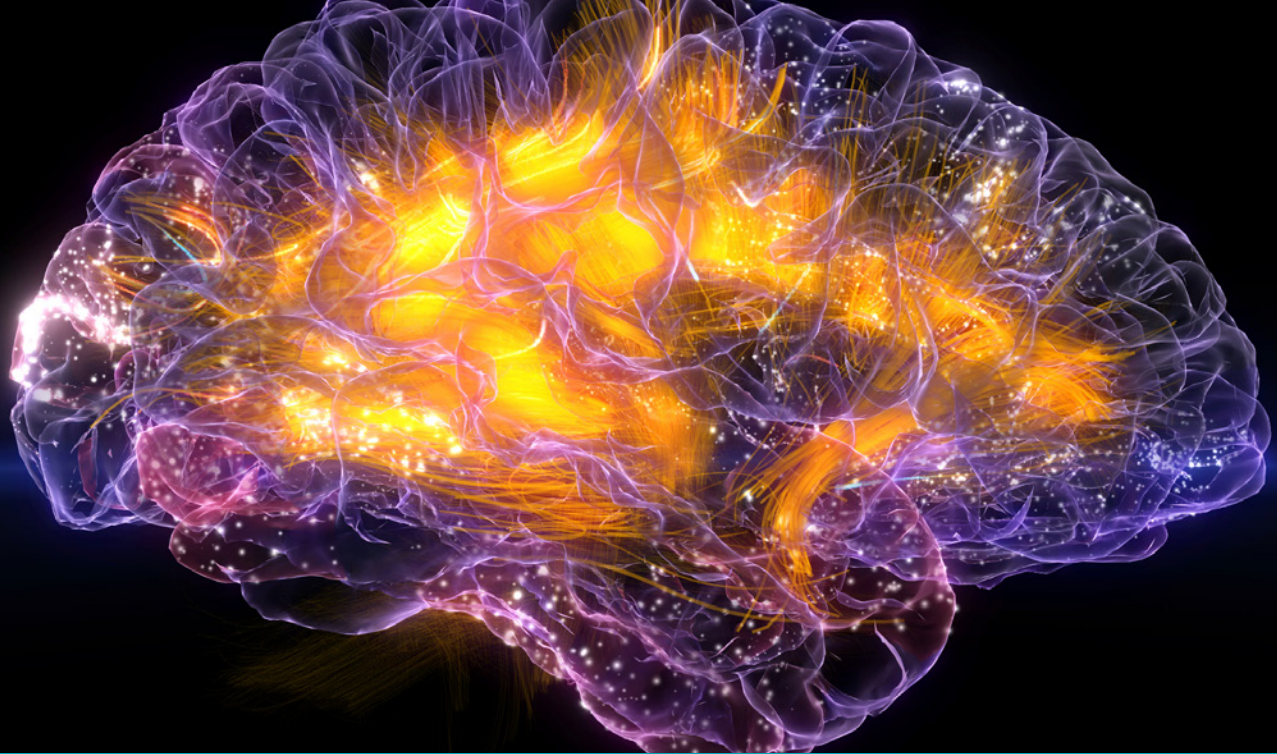


A Healthcare Provider's Guide To Behavioral Variant Frontotemporal Dementia (bvFTD):

Diagnosis, pharmacologic management,
non-pharmacologic management, and
other considerations

This material is provided
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educational resource for
health care providers.



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Diagnosis

Definition

Dementia is a clinical syndrome defined as a cognitive or behavioral decline that leads to an inability to complete daily tasks independently. Dementia has many causes some of which are reversible, such as metabolic disorders, while some are progressive such as Alzheimer disease and behavioral variant frontotemporal dementia (bvFTD). bvFTD is the most common form of frontotemporal dementia (FTD) and is characterized by alterations in social decorum and personal regulation including disinhibition, apathy, overeating, emotional blunting, obsessiveness, repetitive motor behaviors, and impairment in judgment and insight. Along with these behavioral changes, deficits in executive control emerge, and patients have problems with planning, organizing, and generating ideas. Traditional cognitive testing of patients with bvFTD may be relatively normal despite the patient's severe deficits in day-to-day function, and patients typically have little to no insight into the illness.¹

Etiology

bvFTD is one of the most common causes of dementia in people less than 65 years old.² The estimated point prevalence is 15–22/100,000 and incidence 2.7–4.1/100,000. R Approximately 20–50% of individuals with FTD have an affected first-degree relative. Conversely, 50–80% of individuals appear to be the first person with FTD in the family. At autopsy, pathology is variable and could show an accumulation of neurofibrillary tangles made of tau or neurocytoplasmic inclusions made of TDP-43 (TAR DNA-binding protein).

Course

The pace of the symptoms and length of disease can vary dramatically from person to person. In general, each type of FTD follows a pattern where the symptoms seen in the mild stage become more pronounced and disabling over a course of 8–10 years. In early years, a person with bvFTD tends to show marked behavioral changes, apathy and impaired judgment. After several years, the behavioral and cognitive changes become more pronounced, and an MRI will show atrophy. End stage disease includes profound functional impairment with possible language, motor and/or memory difficulty. The Clinical Dementia Rating (CDR) provides a summary of clinical features by dementia stage.³ Patients with FTD have a lower life expectancy compared to that of patients with Alzheimer's disease.^{4,5} Death is not typically due to bvFTD directly, but rather to other complications such as pneumonia, dehydration, urinary tract infection, extensive pressure ulcers, vascular events, or falls and fractures.

Differential Diagnosis

bvFTD is often mistaken for depression, bipolar illness, personality disorder, drug or alcohol dependence, late-onset schizophrenia, Alzheimer's disease, metabolic disorders, vascular dementia, chronic traumatic encephalopathy, structural brain disease, or less commonly with leukoencephalopathies.

Diagnostic Criteria

International consensus research criteria for behavioral variant frontotemporal dementia (bvFTD).⁶

Neurodegenerative disease

The following symptom must be present to meet criteria for bvFTD.

- A. Shows progressive deterioration of behavior and/or cognition by observation or history (as provided by a knowledgeable informant)

Possible bvFTD

Three of the following behavioral/cognitive symptoms must be present to meet criteria. Ascertainment requires that symptoms be persistent or recurrent, rather than single or rare events.

- A. Early behavioral disinhibition
(one of the following must be present)
 - a. Socially inappropriate behavior
 - b. Loss of manners or decorum
 - c. Impulsive, rash or careless actions
- B. Early apathy or inertia
(one of the following must be present)
 - a. Apathy
 - b. Inertia
- C. Early loss of sympathy or empathy
(one of the following must be present)
 - a. Diminished response to other people's needs and feelings
 - b. Diminished social interest, interrelatedness or personal warmth
- D. Early perseverative, stereotyped or compulsive/ritualistic behavior
(one of the following must be present)
 - a. Simple repetitive movements
 - b. Complex, compulsive or ritualistic behaviors
 - c. Stereotypy of speech
- E. Hyperorality and dietary changes
(one of the following must be present)
 - a. Altered food preferences
 - b. Binge eating, increased consumption of alcohol or cigarettes
 - c. Oral exploration or consumption of inedible objects
- F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions
(all of the following must be present)
 - a. Deficits in executive tasks
 - b. Relative sparing of episodic memory
 - c. Relative sparing of visuospatial skills

Probable bvFTD

All of the following symptoms must be present to meet criteria.

- A. Meets criteria for possible bvFTD
- B. Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)
- C. Imaging results consistent with bvFTD (one of the following must be present):
 - a. Frontal and/or anterior temporal atrophy on magnetic resonance imaging (MRI) or computed tomography (CT)
 - b. Frontal and/or anterior temporal hypoperfusion or hypometabolism on positron emission tomography (PET) or single-photon emission computed tomography (SPECT)

Behavioral variant FTD with definite FTLD Pathology

Criterion A and either criterion B or C must be present to meet criteria.

- A. Meets criteria for possible or probable bvFTD
- B. Histopathological evidence of FTLD on biopsy or at post-mortem
- C. Presence of a known pathogenic mutation

Exclusionary criteria for bvFTD

Criteria A and B must be answered negatively for any bvFTD diagnosis. Criterion C can be positive for possible bvFTD but must be negative for probable bvFTD.

- A. Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
- B. Behavioral disturbance is better accounted for by a psychiatric diagnosis
- C. Biomarkers strongly indicative of Alzheimer's disease or other neurodegenerative process

*As a general guideline, "early" refers to symptom presentation within the first three years.

Pharmacologic Management

Medications to Use

Currently, there is no known cure for bvFTD. There are several classes of medications used to treat disease symptoms or improve cognitive function. Selective serotonin re-uptake inhibitors (SSRIs) and atypical antipsychotics may be helpful for managing different behavioral problems.

Review expected and realistic goals of treatment (e.g., treatment is for symptomatic improvement and not a cure or reversal of disease). Expected benefits may be mild improvement in memory function, mood, and alertness. If the patient has vascular disease or mixed dementia, they should receive management and education regarding modification of cardiovascular risk factors.

Medications to Avoid

Medications with strong anticholinergic side effects, such as sedating antihistamines, barbiturates, narcotics, benzodiazepines, gastrointestinal and urinary antispasmodics, central nervous system (CNS) stimulants, muscle relaxants, and tricyclic antidepressants should be avoided. Antipsychotics should be used with caution. If used, carefully evaluate effectiveness of medication and consider discontinuing if there is no improvement in six weeks. Activating medications, such as Ritalin (methylphenidate), should be avoided as they can worsen disinhibition.^{7,8,9}

Non pharmacologic Management

Healthy Lifestyle

There are lifestyle habits that promote health and well-being. Research suggests that the combination of good nutrition, physical activity, and mental and social engagement may provide benefit in



promoting health although more study is needed to determine the actual mechanisms.^{10,11} A heart-healthy diet (lower in sugar and fat and higher in vegetables and fruit) is considered to be good for both the body and the brain. An example is the Mediterranean diet that promotes nutrition based on fruit, vegetables, nuts and grains with limits on consumption of red meat and saturated fats. Physical exercise has been associated with improvement of mood and mobility, and a decrease in the risk for falls.^{12,13} Physical activities that are socially engaging (walking or swimming with a friend and participating in exercise groups) can be especially enjoyable. Engagement in activities that are mentally stimulating (crossword puzzles, sudoku, computer games) is encouraged as long as the activity is enjoyable.

The Alzheimer's Association has more information on tips for maintaining your health.

Sleep

Disrupted sleep can negatively impact memory and thinking, though the mechanisms are not well understood.¹⁴ Components of sleep hygiene include:

- Avoid napping during the day
- Avoid stimulants such as caffeine, nicotine, and alcohol too close to bedtime
- Get regular exercise
- Avoid eating right before sleep
- Ensure adequate exposure to natural light
- Establish a regular relaxing bedtime routine
- Associate your bed with sleep. It's not a good idea to use your bed to watch TV, listen to the radio, or read.

For more details on sleep hygiene, you can refer to the National Sleep Foundation at [sleepfoundation.org/ask-the-expert/sleep-hygiene](https://www.sleepfoundation.org/ask-the-expert/sleep-hygiene).

Other Considerations

Support Resources

Alzheimer's Association: [alz.org](https://www.alz.org)

Family Caregiver Alliance: [caregiver.org](https://www.caregiver.org)

National Institute of Health/National Institute on Aging: [nia.nih.gov/alzheimers](https://www.nia.nih.gov/alzheimers)

The Association for Frontotemporal Degeneration: [theaftd.org](https://www.theaftd.org)

NINDS Frontotemporal Dementia Information Page: [ninds.nih.gov/disorders/picks/picks.htm](https://www.ninds.nih.gov/disorders/picks/picks.htm)

The National Institutes of Health maintains an extensive listing of clinical trials at [clinicaltrials.gov](https://www.clinicaltrials.gov). Academic medical centers may be engaged in research and clinical trials.

Safety

If wandering or getting lost is a concern, refer the patient and family to the [MedicAlert +Alzheimer's Association Safe Return program](https://www.alz.org/medic-alert) (operated by the Alzheimer's Association).

Other strategies for ensuring safety concerns may include door alarms and increased supervision.

Driving

Depending on cognitive and motor findings, the patient may be requested to stop driving, complete test of driving abilities through the Department of Motor Vehicles (DMV), or be referred to a driver's safety course that will assess driving ability. Reporting to the department of motor vehicles should be consistent with state laws. Some states have mandatory reporting requirements: the diagnosis is reported to local health departments who then report to the DMV. Individual state requirements can be found at [dmvusa.com](https://www.dmvusa.com).

Living Situation and Environment

It is important to determine if the patient's residential setting best meets his or her functional and cognitive abilities. Areas of concern may include personal safety (ability to manage medications safely, ability to manage nutritional requirements, ability to manage personal hygiene) and quality of life (activities and engagement that match the person's needs and abilities).

Types of living situations range from living at home alone or living at home with supervision, to board and care, assisted living, or memory care units.

Elder Abuse

Patients with dementia and their caregivers are vulnerable to abuse. Refer to Adult Protective Services (APS) if there is concern for the well-being of the patient or the caregiver.

To locate an APS office in your state, see: napsa-now.org/get-help/help-in-your-area/

Legal Planning

Provide information about advance directives and durable power of attorney while the patient is in the early stages of disease and able to articulate his or her wishes. Make referrals for legal and financial advice, especially if there are concerns about the patient's judgment, decision-making, or vulnerability. A formal evaluation for capacity may be warranted. The Alzheimer's Association provides a [brochure that covers legal planning](#).

Advanced Directives

These documents allow individuals to state their preferences for medical treatments and to select an agent or person to make health care decisions in the event they are unable to do so or if they want someone else to make decisions for them.

Power Of Attorney

A Power of Attorney (POA) is a legal document that gives someone of an individual's choosing the power to act in their place. POAs can be for medical or financial matters.

Living Will

A living will is a written, legal document that spells out medical treatments that an individual would and would not want to be used to keep them alive, as well as other decisions such as pain management or organ donation.

Teaching Video for Providers

An example of a physician telling a patient she has dementia:

alz.org/health-care-professionals/dementia-diagnosis-diagnostic-tests.asp#alzheimers_diagnosis.



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