

What is Frontotemporal Degeneration (bvFTD)?

There is a type of dementia called "frontotemporal" which typically affects patients at a very early age. In this type of dementia, there is no true memory loss in the early stages of the type that is seen in Alzheimer's dementia. Instead, there are changes in personality, ability to concentrate, social skills, motivation and reasoning. Because of their nature, these symptoms are often confused with psychiatric disorders. There are gradual changes in one's customary ways of behaving and responding emotionally to others. Memory, language and visual perception are usually not impaired for the first two years, yet as the disease progresses and spreads to other areas of the brain, they too may become affected. Typically, the disorder affects females more than males.

The symptoms reflect the fact that the brain degeneration is not initially widespread and settles in the parts of the brain that are important for social skills, reasoning, judgement and the ability to take initiative.

When the brains of individuals with frontal lobe dementia are studied after death, the types of microscopic abnormalities that are seen are typically of two kinds. The first type is called Non-specific focal degeneration and the second is labeled Pick's disease. Non-specific focal degeneration accounts for 80% of cases of frontal lobe dementia. It is called "non specific" because there are no abnormal particles that are identifiable-only evidence that brain cells have been eliminated. Pick's disease, which accounts for 20% of cases of frontal lobe dementia, is identified under the microscope by abnormal particles called "Pick bodies", named after the neurologist who first observed them.

How Does it Affect the Brain?

Frontotemporal dementia affects the part of the brain that regulates comportment, insight and reasoning. "Comportment" is a term that refers to social behavior, insight, and "appropriateness" in different social contexts. Normal comportment involves having insight and the ability to recognize what behavior is appropriate in a particular social situation and to adapt one's behavior to the situation. For example, a funeral is a solemn event requiring certain types of behavior and decorum. Similarly, while it may be perfectly natural and acceptable to take one's shoes and socks off at home, it is probably not the thing to do while in a restaurant. Comportment also refers to the style and content of a person's language. Certain types of language are acceptable in some situations or with friends and family, and not acceptable in others.

Insight, an important aspect of comportment, has to do with the ability to "see" oneself as others do. Insight is necessary in order to determine whether one is behaving in a socially acceptable or in a reasonable manner. Insight is also necessary for the patient to recognize his/ her deficits and illness. Changes in comportment may be manifested as "personality" alterations. A generally active, involved person could become apathetic and disinterested. The opposite may also occur. A usually quiet individual may become more outgoing, boisterous and disinhibited. Personality changes can also involve increased irritability, anger and even verbal or physical outbursts toward others (usually the caregiver). Comportment is assessed by observing the patient's behavior throughout the examination and interviewing other people (family and friends) who have information about the patient's "characteristic" behavior.

Individuals with frontotemporal dementia frequently have executive function and reasoning deficits. "Reasoning" refers to mental activities that promote decision-making. Being able to categorize information and to move from one perspective of a problem to another are examples of reasoning. "Executive

functions" is a term that refers to yet another group of mental activities that organize and plan the flow of behavior. A good example of executive functions is what might happen if one were driving a car, talking with the passenger and suddenly having to respond to a child running into traffic. The ability to handle all the stimulation and to quickly plan a course of action is accomplished via executive functions. Individuals with frontal lobe dementia often lack flexibility in thinking and are unable to carry a project through to completion. Failure of executive functions may increase safety risk since they may not be able to plan appropriate actions or inhibit inappropriate actions.

Signs & Symptoms of bvFTD

- Impairments in social skills
 - inappropriate or bizarre social behavior (e.g., eating with one's fingers in public, doing sit-ups in a public restroom, being overly familiar with strangers)
 - "loosening" of normal social restraints (e.g., using obscene language or making inappropriate sexual remarks)
- Change in activity level
 - apathy, withdrawal, loss of interest, lack of motivation, and initiative which may appear to be depression but the patient does not experience sad feelings.
 - in some instances there is an increase in purposeless activity (e.g., pacing, constant cleaning) or agitation.
- Decreased Judgment
 - impairments in financial decision- making (e.g., impulsive spending)
 - difficulty recognizing consequences of behavior
 - lack of appreciation for threats to safety (e.g., inviting strangers into home)
- Changes in personal habits
 - lack of concern over personal appearance
 - irresponsibility
 - compulsiveness (need to carry out repeated actions that are inappropriate or not relevant to the situation at hand).
- Alterations in personality and mood
 - increased irritability, decreased ability to tolerate frustration
- Changes in one's customary emotional responsiveness
 - a lack of sympathy or compassion in someone who was typically responsive to others' distress
 - heightened emotionality in someone who was typically less emotionally responsive

Persons with this form of dementia may look like they have problems in almost all areas of mental function. This is because all mental activity requires attention, concentration and the ability to organize information, all of which are impaired in frontal lobe dementia. Careful testing, however, usually shows that most of the problems stem from a lack of persistence and increased inertia.

Diagnosis & Treatment

The absolute diagnosis of Alzheimer's disease can only be made at autopsy. However, physicians at specialized centers can diagnose Alzheimer's disease with 90 percent certainty based on clinical information. To make the diagnosis the following may need to be conducted:

- A medical history and neurological exam
- Neuropsychological testing
- Neuropsychological testing involves a careful analysis of a person's memory, problem solving, language, attention, and visuospatial ability.
- Basic blood tests
- Blood tests may be used to help exclude other causes of memory difficulties. For example, a person with a thyroid disorder or a vitamin deficiency may have problems with his or her memory.
- Brain scans
- A brain scan such as an MRI or a CT scan may need to be done in certain patients to detect brain tumors or strokes. These disorders may cause memory problems.

Fortunately, there are steps that can be taken to provide a secure environment for the diagnosed person and obtain help for family:

- Obtain a psychiatric evaluation from an individual with experience treating people with dementia. Certain medications can help with behavior problems such as agitation and hostility.
- Share information with family and friends. This will help them better understand the patient's behavior and provide an opportunity for them to offer the diagnosed person and their family some support and respite.
- Encourage the person to attend an early stage support group. Even if the support group is geared toward the person with early Alzheimer's disease, much information will also be relevant to Frontal Lobe Dementia.
- Meet with an attorney or financial consultant. Make sure Durable Power of Attorney forms have been completed for both health care and finances. Give copies to your doctor. An "elderlaw" attorney who is well-versed in these issues is still an appropriate choice to help you draft these documents or you may obtain the forms at many stationary stores and complete them on your own.
- Attend a caregiver support group. Listening to others who are going through similar experiences can be very comforting. They may also aid you in developing new caregiver techniques and learn about different resources within your community.
- Try to remain physically and mentally healthy. Be sure to get regular health check-ups for both the diagnosed person and family. Exercise and eat nutritious meals. Build in time for things that allow you to rejuvenate.
- Obtain a driving evaluation: Contact your local Alzheimer's Association for the driving evaluation program near you.

Living with bvFTD

The psychological, social, family and financial issues that affect individuals with frontotemporal dementia are drastically different from those that affect individuals with Alzheimer's type dementia. When dementia occurs earlier in life, issues such as working, teenage children and financial stress are different from the issues dealt with by individuals who are older and most likely retired. Planning for the family's financial security and for the education of children becomes a difficult prospect when an individual is faced with a dementing illness in the prime of his/her working career. The nature of the symptoms themselves are often embarrassing to family members and there may be loss of friends and other sources of social support. Finally, most adult day programs and residential care facilities are not equipped to address the

special needs of the younger patient, especially if the behavioral symptoms are difficult to manage. As more is known about the disease, more policy changes may come into effect. Some residential care and adult day programs are recognizing the needs of the younger dementia patient and are beginning to offer services to meet their needs. Before making any decisions, it is best to investigate your options.

Depending on severity, a patient with impaired comportment may not be able to manage their daily activities without supervision. They may be at risk for harming themselves or being victimized because they would not be able to recognize their limitations or use proper judgement. Driving is usually unsafe for persons with this diagnosis.

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Resources for bvFTD

The Association for Frontotemporal Dementias

www.ftd-picks.org

info@ftd-picks.org

If the patient is working and needs to file for disability, it is best to speak to their employer as well as the local social security office. Disability benefits are usually obtained as long as your impairment does not medically improve and you cannot work.

Social Security's Toll-Free Number: 1-800-772-1213

Internet address: www.ssa.gov

You can obtain more information on resources from your local Alzheimer's Association.

The Alzheimer's Association, national number: 1-800-272-3900

Internet address: www.alz.org

An "elder law" attorney can assist you with legal and financial planning.
The National Academy of ElderLaw Attorneys: 1-520-881-4005
Internet address: www.naela.org/

The Pick's Disease Support Group www.pdsq.org.uk/index.htm
For carers of frontotemporal dementia: Pick's Disease, Frontal Lobe Degeneration, Dementia with Lewy Bodies, Corticobasal Degeneration and Alcohol Related Dementia.

National Organization for Rare Disorders (NORD)
Phone: 800-447-6673
www.rarediseases.org

Alzheimer's Disease Education and Referral Center
Phone: 1-800-438-4380
www.alzheimers.org/

NIH/National Institute of Neurological Disorders and Stroke
(800) 352-9424
www.ninds.nih.gov

NIH/National Institute on Aging
(800) 222-2225
www.nih.gov/nia.htm