

Frontotemporal dementia (FTD)

A topic in the Alzheimer's Association® series on understanding dementia.

About dementia

Dementia is a condition in which a person has significant difficulty with daily functioning because of problems with thinking and memory. Dementia is not a single disease; it's an overall term — like heart disease — that covers a wide range of specific medical conditions, including Alzheimer's disease. Disorders grouped under the general term “dementia” are caused by abnormal brain changes. These changes trigger a decline in thinking skills, also known as cognitive abilities, severe enough to impair daily life and independent function. They also affect behavior, feelings and relationships.

Brain changes that cause dementia may be temporary, but they are most often permanent and worsen over time, leading to increasing disability and a shortened life span. Survival can vary widely, depending on such factors as the cause of the dementia, age at diagnosis and coexisting health conditions.

Frontotemporal dementia or frontotemporal degenerations

Frontotemporal dementia refers to a group of disorders that cause progressive nerve cell loss in the frontal and temporal lobes of the brain. The nerve cell damage leads to loss of function in these brain regions, which can variably cause deterioration in behavior and personality, language disturbances or alterations in muscle and motor functions.

There are a number of different diseases that cause frontotemporal degenerations. The two most prominent are a group of brain disorders involving tau protein and others involving TDP43 protein. For reasons that are not yet known, these two groups have a predilection for the frontal and temporal lobes.

Behavior variant frontotemporal dementia

Behavior variant frontotemporal dementia (bvFTD) is a condition characterized by prominent changes in personality, interpersonal relationships and conduct that often occurs in people in their 50s and 60s but can develop as early as their 20s or as late as their 80s. The most common behaviors are caused by the degeneration of neuronal cells in the frontotemporal area of the brain. In bvFTD, the nerve cell loss is most prominent in areas that control conduct, judgment, understanding of how other people feel (empathy) and foresight, among other abilities.

BvFTD is suspected as a diagnosis when a person who previously exhibited normal behavior and judgment shows changes in personality, interpersonal skills and other daily behaviors. The changes in personality might include disinterest in prior pastimes or family affairs. BvFTD could lead to loss of interpersonal skills that may result in more socially inappropriate activities, such as making inappropriate comments or acting in demeaning, rude

or immodest manners. In addition, a loss of empathy is common and catastrophically poor judgment could lead to major financial or personal crises.

Primary progressive aphasia

Primary progressive aphasia (PPA) is the other major form of frontotemporal degeneration. Aphasia refers to a disorder of language that can involve problems with speaking, writing or comprehension. The “primary progressive” refers to the usual way in which PPA occurs, namely first and foremost as a disorder of language. PPA normally develops before the age of 65 but can occur in late life also. In contrast to bvFTD, the brain degeneration that occurs in PPA tends to be much more limited to the left hemisphere of the brain, where, for most people, language functions in the brain are located. Different speech and language difficulties arise depending on where the brunt of the degeneration occurs, which could include the left frontal, temporal or parietal lobes.

The two most distinctive PPA subtypes are one called nonfluent/agrammatic variant of PPA, in which speaking is very hesitant, labored, stuttered and telegraphic. The other distinctive PPA subtype is called the semantic variant of PPA, where the affected individual loses the ability to understand or formulate words in a spoken sentence.

Disturbances of motor (movement or muscle) function in frontotemporal degenerations

Three disorders on the frontotemporal degeneration spectrum produce changes in muscle or motor functions with or without behavior changes (such as in bvFTD) or problems with language (like those in PPA). The first, amyotrophic lateral sclerosis (ALS, also known as motor neuron disease or Lou Gehrig’s disease), is a disorder of muscle weakness and wasting. The second, corticobasal syndrome, is a disorder in which the arms and legs become uncoordinated and stiff. The third, progressive supranuclear palsy (PSP), is a disorder in which there are problems with eye movement, as well as muscle stiffness, difficulty walking and changes in posture.

These disorders share the same abnormal changes in the brain that occur in bvFTD and PPA except that the nerve cell degenerative changes are primarily in parts of the brain that affect the motor nerve cells responsible for coordinating muscle movement rather than behavior or language.

Prevalence of frontotemporal degenerations

Both bvFTD and PPA are far less common than Alzheimer’s disease in individuals over age 65. However, in the 45 to 65 age range, bvFTD and PPA are nearly as common as younger-onset Alzheimer’s disease. Only rough estimates are available, but there may be 50,000 to 60,000 people with bvFTD or PPA in the United States — the vast majority between ages 45 and 65. It is rare that these disorders occur outside of this age range.

In the United States, there are approximately 30,000 cases of ALS and approximately 20,000 of PSP.

Diagnosis

Diagnoses of bvFTD and PPA are based on expert evaluation by a physician who is familiar with these disorders. The patient's symptoms and results from neurological examinations are the core of the diagnosis. Brain scans such as magnetic resonance imaging (MRI) and glucose positron emission scans are helpful diagnostic tests but must be evaluated together with the patient's medical history and neurological examination. Occasionally a psychiatric evaluation will also be ordered as part of the diagnostic process, such as when it is not clear if changes in behavior are due to depression or another psychiatric disturbance.

Since a decline in language abilities is the primary symptom of PPA, it is important to determine which components of language use are most affected, how severely affected they are and what can be done to improve communication. A speech-language pathologist evaluates different aspects of language in detail and can make recommendations for strategies to improve communication. Family members should be included in the treatment sessions to educate them about how to facilitate communication.

Causes and risks

Frontotemporal degenerations are inherited in about a third of all cases. Genetic counseling and testing are available now for individuals with a family history of frontotemporal degenerations. Family history is the only known risk factor for any of the frontotemporal degenerations.

Outcomes

Frontotemporal degenerations worsen over time, but speed of decline varies from person to person. Individuals with FTD may experience muscle weakness and coordination problems for many years, often requiring use of a wheelchair or confining them bedridden. These muscle issues can cause problems with swallowing, chewing, moving and controlling bladder and/or bowel function. As a result of their vulnerable physical state, many individuals with frontotemporal degenerations die from complications with skin, urinary tract and/or lung infections.

Treatment

No specific treatments exist for any of the frontotemporal degeneration disorders. However, medications can reduce agitation, irritability and/or depression, and help improve overall quality of life. Speech therapy may be beneficial to individuals with PPA, as it can focus directly on the language skills that are impaired.

Resources

Association for Frontotemporal Degeneration (AFTD)

theaftd.org
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