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Understanding Frontotemporal Dementia

Frontotemporal dementia (FTD) or frontotemporal degenerations refers to a group of disorders caused by progressive nerve cell loss in the brain's frontal lobes (the areas behind your forehead) or its temporal lobes (the regions behind your ears).

The nerve cell damage caused by frontotemporal dementia leads to loss of function in these brain regions, which variably cause deterioration in behavior and personality, language disturbances, or alterations in muscle or motor functions.

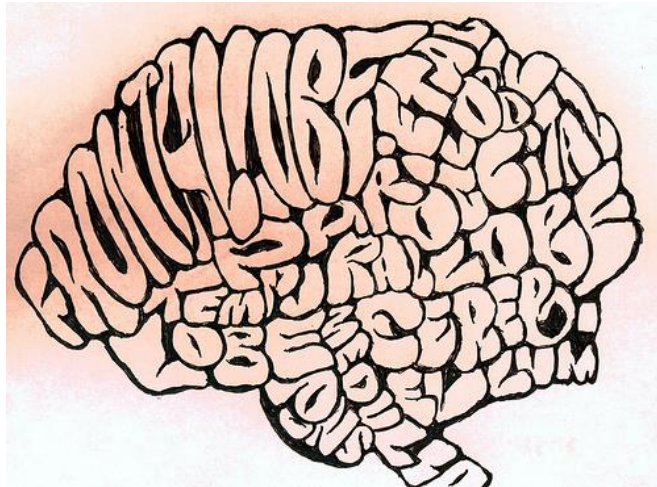
There are a number of different diseases that cause frontotemporal degenerations. The two most prominent are 1) a group of brain disorders involving the protein tau and 2) a group of brain disorders involving the protein called TDP43. For reasons that are not yet known, these two groups have a preference for attacking a particular kind of brain cells, the spindle neurons, which are found in the fronto-insular cortex of the brain.

FTD used to be called Pick's disease after Arnold Pick, a physician who in 1892 first described a patient with distinct symptoms affecting language. Some doctors still use the term "Pick's disease", mostly when referring to the behavior variant of FTD. Other terms you may see used to describe FTD include frontotemporal disorders, frontotemporal degenerations and frontal lobe disorders.

The disorders grouped under FTD fall into three subtypes.

Types of FTD

Behavior variant frontotemporal dementia (bvFTD). This condition is characterized by prominent changes in personality, interpersonal relationships and conduct that often occur in people in their 50s and 60s, but can develop as early as their 20s or as late as their 80s. In



bvFTD, the nerve cell loss is most prominent in areas that control conduct, judgment, empathy and foresight, among other abilities.

Caregivers of bvFTD patients often report selfish behaviors (i.e. hoarding candy and hiding treats from family members), preceding the diagnosis by many years, sometimes decades. Overtime, such behaviors may have been cause for loss of interpersonal relationships and isolation. As with in all types of FTD, it is believed that the loss of neurons in bvFTD starts up to 20 years before symptoms become noticeable, which may account for these reports.

Patients with bvFTD may act in socially inappropriate manners, and generally have a difficult time controlling anger, frustration and sexual urges. Caregivers must pay extra attention to safety measures (i.e. removing guns and sharp objects from the home) and have emergency plans for responding to any event of extreme anger or agitation.

Primary progressive aphasia (PPA). This is the second major form of frontotemporal degeneration that affects language skills, speaking, writing and comprehension. PPA normally comes on in midlife, before age 65, but can occur in late life also. The two most distinctive forms of PPA have somewhat different symptoms:



Tips

Key Differences Between FTD and Alzheimer's

Age at diagnosis. Most people with FTD are diagnosed in their 40s and early 60s. Alzheimer's, on the other hand, grows more common with increasing age.

Memory loss tends to be a more prominent symptom in early Alzheimer's than in early FTD, although advanced FTD often causes memory loss in addition to its more characteristic effects on behavior and language.

Behavior changes are often the first noticeable symptoms in bvFTD, the most common form of FTD. Behavior changes are also common as Alzheimer's progresses, but they tend to occur later in the disease.

Problems with spatial orientation — for example, getting lost in familiar places — are more common in Alzheimer's than in FTD.

Problems with speech. Although people with Alzheimer's may have trouble thinking of the right word or remembering names, they tend to have less difficulty making sense when they speak, understanding the speech of others, or reading than those with FTD.

Hallucinations and delusions are relatively common as Alzheimer's progresses, but relatively uncommon in FTD.

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- ◆ In **semantic variant of PPA**, individuals lose the ability to understand or formulate words in a spoken sentence.
- ◆ In **nonfluent/agrammatic variant of PPA**, a person's speaking is very hesitant, labored or ungrammatical.

Disturbances of motor (movement or muscle) function. There are three disorders that are a part of the frontotemporal degeneration spectrum that produce changes in muscle or motor functions with or without behavior (bvFTD) or language (PPA) problems.

Amyotrophic lateral sclerosis (ALS), which causes muscle weakness or wasting. ALS is a motor neuron disease also known as Lou Gehrig's disease.

Corticobasal syndrome, which causes arms and legs to become uncoordinated or stiff.

Progressive supranuclear palsy (PSP), which causes muscle stiffness, difficulty walking and changes in posture. It also affects eye movements.

Both bvFTD and PPA are far less common than Alzheimer's disease in those over age 65 years. However, in the 45 to 65 age range, bvFTD and PPA are nearly as common as younger-onset Alzheimer's. Only rough estimates are available, but there may be 50,000 to 60,000 people with bvFTD and PPA in the United States, the majority of whom are between 45 and 65 years of age.

Causes and Risks

The biggest risk factor for any frontotemporal degenerations is a family history or a similar disorder. Frontotemporal degenerations are inherited in about 40 to 50% of all cases. There are 5 genes that have been identified as determining of the disease but they do not account for all cases of FTD, so it is possible that more

related genes will be found in the future. Genetic counseling and testing is available for individuals with family histories of frontotemporal degenerations.

There is also a connection between FTD and brain injuries, as well as thyroid disease. Studies have found that head trauma was associated with a 3.3-fold higher risk for FTD, and thyroid disease with a 2.5-fold higher risk.

Diagnosis

The diagnosis of bvFTD and PPA are based on expert evaluation by a doctor who is familiar with these disorders. The type of problems experienced by the patient and the results of neurological exams are the core of the diagnosis. Brain scans such as magnetic resonance imaging (MRI) and Positron emission tomography (PET-scan) can show the atrophy in the frontal and temporal lobes of the brain. However, all test results must be interpreted in the context of the patient's history and neurological exam.

Treatment and Outcomes

There are no specific treatments for any of the frontotemporal subtypes. There are medications that can reduce agitation, irritability and/or depression. These treatments should be used to help improve quality of life.

FTD inevitably gets worse over time and the speed of decline differs from person to person. For many years, individuals with FTD show muscle weakness and coordination problems, leaving them wheelchair or bedbound. These muscle issues can cause problems swallowing, chewing, moving and controlling bladder and/or bowels. Eventually people with frontotemporal degenerations die because of the physical changes that can cause skin, urinary tract and/or lung infections.



Tools:

The Association for Frontotemporal Degeneration (AFTD) is a nonprofit organization that provides information, education and support to those affected by FTD and their caregivers. Call AFTD at 866.507.7222.

The Alzheimer's Association can help you learn more about Alzheimer's and other dementias, and help you find local support services. Call our 24/7 Helpline at 800.272.3900. **Social Security Administration (SSA)** has a "compassionate allowance" program in which workers diagnosed with Pick's disease, PPA or ALS can qualify for Social Security disability benefits. You can also call the SSA at 800.772.1213.