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 lifespan. Survival can vary widely, depending on such factors as the cause of the dementia, age at diagnosis and coexisting health conditions.

Frontotemporal Dementia

Frontotemporal dementia (FTD) is a group of disorders caused by progressive nerve cell degeneration in the brain's frontal lobes (the areas behind your forehead) or its temporal lobes (the regions behind your ears). This cell damage leads to tissue shrinkage and reduced function in these brain regions, which control planning and judgment, emotions, speaking and understanding speech, and certain types of movement.

FTD includes a range of specific conditions with different core symptoms. But there's significant symptom overlap, especially as these disorders progress. The disorders grouped under FTD fall into three broad categories (noted below under Symptoms). Scientists have identified a range of microscopic brain abnormalities implicated in FTD. The overall term for the brain cell damage and tissue shrinkage associated with FTD is frontotemporal lobar degeneration (FTLD).

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." Other terms you may see used to describe FTD include frontotemporal disorders, frontotemporal degeneration and frontal lobe disorders.

Prevalence

FTD, once considered rare, is now thought to account for up to 10 to 15 percent of all dementia cases. It's still believed to be less common than Alzheimer's disease, vascular dementia or Lewy body dementia. In those younger than age 65, FTD may account for up to 20 to 50 percent of dementia cases. People usually develop FTD in their 50s or early 60s, making the disorder relatively more common in this younger age group.

Symptoms

Initially, the three main categories of FTD tend to cause different core symptoms based on the first parts of the brain's frontal or temporal lobes they affect. But there's increasing overlap in symptoms as these disorders progress.

Core symptoms of each type of FTD include the following:

- **Behavioral variant frontotemporal dementia (bvFTD)** takes its greatest toll on personality and behavior. It may begin with subtle changes that may be mistaken for depression. As bvFTD progresses people often develop disinhibition, a striking loss of restraint in personal relations and social life.
- **Primary progressive aphasia (PPA)** chiefly affects language skills in early stages, but often also affects behavior as it advances. The two chief forms of PPA have somewhat different symptoms:
    - In **semantic dementia**, people speak easily, but their words convey less and less meaning. They tend to use broad, generic terms, such as "animal" when they mean "cat." Language comprehension also declines.
    - In **progressive nonfluent aphasia**, people lose their ability to generate words easily, and their speech becomes halting, "tongue-tied" and ungrammatical. Ability to read and write may also be impaired.
- **FTD movement disorders** primarily affect certain involuntary, automatic muscle functions. These disorders may also impair language and behavior. The two primary FTD movement disorders are:
    - **Corticobasal degeneration (CBD),** which causes shakiness, lack of coordination, and muscle rigidity and spasms.
    - **Progressive supranuclear palsy (PSP),** which causes walking and balance problems, frequent falls and muscle stiffness, especially in the neck and upper body. It also affects eye movements.

Diagnosis

There is no single test — or any combination of tests — that can conclusively diagnose FTD. FTD is a "clinical" diagnosis representing a doctor's best professional judgment about the reason for a person's symptoms. Magnetic resonance imaging (MRI) often plays a key role in diagnosis because it can detect the shrinkage in the brain's frontal and temporal lobes that's a hallmark of FTD.

In some cases, it may be hard to distinguish FTD from Alzheimer's disease. Age at diagnosis may offer one clue. Most people with FTD are diagnosed in their 50s and early 60s. Only about 10 percent are diagnosed over age 70. Alzheimer's, on the other hand, is more common with increasing age. In the future, tests to detect specific protein abnormalities linked to Alzheimer's and FTD may help clarify the diagnosis.

Causes and Risk Factors

Researchers have identified abnormal deposits of several proteins inside the brain cells of those who died with FTD. Scientists have not yet learned what causes these protein abnormalities or

solved the mystery of why the damage associated with FTD targets the brain's frontal and temporal lobes.

The only known risk factor for FTD is a family history of the disease. Scientists have found several genes linked to FTD.

Outcomes

FTD inevitably gets worse, usually over several years. In advanced FTD, people typically become mute and bedbound. Like other types of dementia, FTD shortens lifespan. Studies suggest that most people with FTD survive an average of six to eight years**,** but survival can range from two to 20 years.

Treatment

Current FTD treatment focuses on managing symptoms, primarily those affecting behavior. Emerging insights into specific protein abnormalities associated with FTD may identify targets for new treatments aimed at underlying disease processes.

Antidepressants and antipsychotic drugs are the chief medications used to treat behavioral FTD symptoms. None of these drugs have been approved by the U.S. Food and Drug Administration (FDA) for use in FTD.

Learn More

For more information on frontotemporal dementia and other topics in the Alzheimer's Association series on understanding dementia, visit www.alz.org, or call our toll-free, 24/7 Helpline at 800.272.3900.

The Alzheimer's Association is the world's leading voluntary health organization in Alzheimer's care, support and research.