FTD OVERVIEW

FTD, or frontotemporal degeneration, represents a group of brain disorders caused by the degeneration or shrinking of the frontal and/or temporal lobes of the brain. It is also frequently referred to as frontotemporal dementia, frontotemporal lobar degeneration, or Pick's disease.

FTD is a progressive disease; as time passes, symptoms are seen more frequently, and grow more intense. The length of progression varies from 2 to over 20 years. Often, the first indications of FTD are overlooked or dismissed as someone simply misspeaking or acting out of character, maybe because they are having a bad day. But as one's FTD progresses, it becomes increasingly difficult to plan or organize activities, behave appropriately in social or work settings, and care for oneself, resulting in increasing dependency. Changes to one's behavior and personality, along with difficulties in language and movement, are FTD's hallmark symptoms – unlike Alzheimer's disease, which is primarily characterized by memory loss.

As FTD progresses, it predisposes an individual to physical complications such as pneumonia, infection, or injury from a fall, with an average life expectancy of 7 to 13 years after the start of symptoms.

FTD is often misdiagnosed as a different dementia (such as Alzheimer's), some other type of neurological disorder, or a psychiatric problem such as depression. AFTD has created diagnostic checklists to help families get an accurate diagnosis by identifying red flags to physicians. Diagnosing FTD requires a thorough history, verified by a care partner or caregiver, and a neurological examination.

FTD Subtypes (Ordered by Prevalence)

BEHAVIORAL VARIANT FTD (bvFTD)

Behavioral variant FTD is the most common form of FTD, responsible for about half of all cases. BvFTD is also frequently referred to as frontotemporal dementia or Pick's disease, and is identified by personality changes, apathy, and a progressive decline in socially appropriate behavior, judgment, self-control, and empathy. People with bvFTD typically do not recognize the changes in their own behavior, or exhibit awareness or concern for how their behavior affects others.

FTD OVERVIEW

PRIMARY PROGRESSIVE APHASIA (PPA)

PPA is the second most common type of FTD, characterized by a gradual loss of the ability to speak, read, write, or understand what others are saying. Experts further organize PPA into three groups based on which language skills are most affected:

- **Nonfluent/agrammatic variant PPA**, or progressive nonfluent aphasia, makes producing speech difficult. While the person diagnosed can still recall the meanings of individual words, their speech becomes slow and effortful. They may omit words, use incorrect word endings, and/or mix up the order of words in sentences.
- **Semantic variant PPA**, or semantic dementia, causes one to "lose" the meanings of words and struggle to recall the names of objects. Speech becomes vague and difficult to understand because many words are omitted or substituted.
- **Logopenic variant PPA** makes it difficult to remember words while speaking, causing slow and hesitant speech, although the understanding of words' meanings remains intact. Persons diagnosed may also have short-term memory problems and repeat phrases and sentences.

ALS AND FRONTOTEMPORAL DEGENERATION

Also known as FTD with motor neuron disease (MND), this subtype causes difficulty with walking, standing, using one's hands, speaking, swallowing, or breathing. Other motor symptoms may include muscle weakness, atrophy, twitching, or slurred speech. BvFTD symptoms (such as changes in behavior, personality, and language skills) are often noticed first. Less commonly, symptoms of the nonfluent agrammatic and semantic variants of PPA are seen.

There are several gene variants known to cause FTD, ALS, and ALS with FTD, but the most common gene is *C9orf72*. Since both FTD and ALS can occur in the same person, FTD and ALS researchers often collaborate in their work.

CORTICOBASAL SYNDROME (CBS)

In addition to the frontal and temporal lobes of the brain, several regions deeper in the brain that initiate, control, and coordinate movement are affected in CBS, which presents as a decline in motor function very similar that found in Parkinson's disease (and is sometimes referred to as atypical Parkinsonism). Movement difficulties in CBS often begin on one side of the body but will spread to the other side. As the disease progresses, changes in behavior and language skills common to bvFTD and PPA may appear.

PROGRESSIVE SUPRANUCLEAR PALSY (PSP)

PSP symptoms also resemble those seen in Parkinson's disease. Its earliest motor symptoms are muscular stiffness in the neck and trunk, along with poor balance and more frequent falls. A red flag for PSP is an inability to easily point one's eyes upward; eye movement becomes even more restricted from there, and even opening and closing one's eyes becomes difficult. PSP can also affect overall coordination, along with movement of the mouth, tongue, and throat. In addition to motor symptoms, people with PSP may exhibit changes in behavior and language skills common to bvFTD and PPA, particularly as the disease progresses.

FTD OVERVIEW

FTD & Genetics

When a person is diagnosed with FTD, their relatives may worry that they could develop it themselves. AFTD strongly recommends talking to a genetic counselor to help you assess your risk by evaluating your personal and family health history. Genetic counselors are trained to guide you through this process and provide support as you consider genetic testing.

How does FTD develop? There are several ways that FTD cases can be characterized:

- **Sporadic FTD** – In approximately 60% of people diagnosed with FTD, there is no family history of FTD or any other neurological condition. We refer to FTD cases where the cause is not clearly understood as "apparently sporadic FTD." A genetic evaluation should still be considered, as a genetic cause of FTD can be identified in a small percentage of apparently sporadic FTD cases.
- **Familial FTD** – Approximately 40% of people diagnosed with FTD have a family history of one or more blood relatives diagnosed with FTD or a related condition (such as ALS), a mental health condition like depression or anxiety, progressive challenges with language or movement, or another dementia.
- **Genetic FTD** – In a portion of those with familial FTD, a genetic variant (or mutation) can be identified as the cause: an affected parent has passed a genetic variant associated with FTD to their child. All known genetic forms of FTD are inherited in an autosomal dominant manner, meaning the child of a person with FTD has a 50% chance of inheriting the FTD-causing variant. Variants in more than a dozen genes can cause FTD; however, the most common genes are *C9orf72*, *GRN*, and *MAPT*.

ASK FOR A REFERRAL TO A GENETIC COUNSELOR

Your neurologist or the AFTD HelpLine can assist you in finding a genetic counselor or other healthcare professional experienced in the genetics of adult neurological conditions.

FTD OVERVIEW

Life With FTD

There are currently no approved treatments for FTD, but steps can be taken to manage symptoms.

FOLLOW A DAILY ROUTINE

Many people with FTD, as well as their care partners, benefit from following a regular daily routine. Predictable patterns and activities – meals, household tasks, physical activity, hobbies, social interaction, spiritual development, a regular sleep schedule – provide an important framework for both the person with FTD and the caregiver.

Since the course of FTD is unpredictable, both care partners and persons diagnosed should try to remain flexible. While you may not be able to do things as quickly or easily as before, elements of your routine can be adapted to your changing needs and still provide enjoyment. As symptoms progress, however, you may need to discontinue some parts of your routine.

ENGAGE IN ACTIVITIES

It is also important for people with FTD to stay engaged in activities they find fun and stimulating. An activity can be as simple as listening to music or watching the birds outside, as long as it brings enjoyment. If competitive poker or bridge was a favorite social hobby, playing a more casual or simpler version (with fewer rules, if needed) can engage the person, connect them with their past, and provide a meaningful way to interact with others. However, if an activity stops being enjoyable and starts causing frustration as one's abilities change, it is time to reevaluate and adjust.

ADJUST YOUR CAREGIVING MINDSET

Since people with FTD often cannot be expected to change their behavior, care partners and caregivers must adjust their own expectations when it comes to managing symptoms. Confronting a person diagnosed and trying to correct their actions after the fact is usually less successful than trying to prevent these behaviors before they happen. Try to notice what triggers certain symptoms (such as aggressive or disinhibited behavior) and avoid them if possible. Such triggers could include: certain activities of daily living, the presence of other individuals, certain times of day, and specific environmental factors (noisy and/or unfamiliar places, other visual triggers).

AFTD Is Here to Help

You don't have to face the FTD journey without help. AFTD has support groups and educational resources you can use to teach yourself and others about this disease. Visit our website at **theaftd.org**, or contact the AFTD HelpLine at **866.507.7222** or **info@theaftd.org** to learn more.

The Association for Frontotemporal Degeneration
theaftd.org | HelpLine: 1.866.507.7222 | info@theaftd.org