Ochsner Health

Ochsner Neurocognitive Program

Understanding Frontotemporal Lobar Degeneration

Frontotemporal Lobar Degeneration (FTLD) is a group of neurodegenerative conditions that primarily affect the **frontal** and **temporal** lobes of the brain. These regions are responsible for behavior, personality, judgment, language, and aspects of movement. Because these systems are affected early, FTLD often looks very different from Alzheimer's disease.

FTLD is one of the most common causes of early-onset dementia (before age 65), though it can occur later in life.

What Makes FTLD Different from Alzheimer's Disease?

Unlike Alzheimer's disease, **memory is often relatively preserved early**. Initial symptoms more often include:

- Changes in personality or social behavior
- Poor judgment, impulsivity, or apathy
- Loss of empathy or emotional awareness
- Changes in speech or word understanding
- In some forms, changes in movement or balance

These features are often mistaken for stress, depression, or psychiatric illness early on.

Why FTLD Looks Different in Different People

FTLD is not a single disease. Symptoms depend on which brain networks are affected and may involve behavior, language, or movement systems. This explains why people with FTLD can look very different from one another.

What Causes FTLD?

FTLD is caused by abnormal buildup of proteins in brain cells, most commonly tau or TDP-43, which disrupt brain communication over time. Some cases are inherited, but many occur without a family history.

What to Expect

FTLD is progressive, but the **rate of change varies widely**. Periods of stability are common. While there is no cure, care focuses on safety, communication support, daily structure, and caregiver support.

What This Means

FTLD primarily affects behavior, language, and judgment, often before memory changes appear. Understanding this helps families recognize these changes as neurologic in origin and supports earlier, more appropriate care and planning.

Key Points

- FTLD affects the frontal and temporal lobes
- Often mistaken for psychiatric or stress-related conditions
- Symptoms vary based on affected brain networks
- Caused by abnormal brain protein buildup (commonly tau or TDP-43)
- There is no cure for FTlD
- Progressive, with highly variable pace
- Care focuses on safety, structure, and support

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Why Frontotemporal Lobar Degeneration Presents Differently

One Name. Different Diseases. Different Patterns.

Frontotemporal lobar degeneration (FTLD) is a **group of related brain diseases**, not a single condition.

What makes FTLD look different from person to person is **which part of the frontal or temporal lobes is affected first and most strongly**. These brain systems control behavior, language, executive skills, and movement. Because of this, the **first and most noticeable symptoms can vary**, even though the underlying disease process is related.

Over time, more than one brain system is often involved.

Behavior & Personality

Behavioral variant
- Acting impulsively or without social restraint
- Reduced empathy, warmth, or concern for others
- Apathy, rigidity, or socially inappropriate behavior

Language & Comprehension

Semantic variant
- Trouble understanding word meanings or concepts
- Difficulty naming objects or people
- Fluent speech that lacks clear meaning

Language & Production

Agrammatic/Nonfluent variant
- Speech becomes slow, effortful, or halting
- Difficulty forming sentences or getting words out
- Speech may sound strained or grammatically incorrect

Movement of Limbs/Body

Corticobasal variant
- Stiffness, clumsiness, or slowed movement
- Difficulty using one hand for skilled tasks
- Balance problems or unexplained falls

Movement of Head/Eyes

Progressive Supranuclear palsy variant
- Difficulty moving the eyes up or down
- Reduced facial expression or blinking
- Balance or walking problems related to eye movement

Key Points

- FTLD includes several related but distinct conditions.
- Symptoms differ based on which brain areas are affected.
- Early changes may involve behavior, language, or movement.
- These symptoms reflect brain changes, not choice or personality.
- Recognizing patterns helps families and clinicians plan care.

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What Causes Frontotemporal Lobar Degeneration

Frontotemporal lobar degeneration (FTLD) is a **group of related brain diseases**, not a single condition. It is usually caused by **abnormal buildup of proteins in brain cells**, which disrupt communication over time .

The Most Common Causes of FTLD

- **TDP-43 (Most Common)**
- **Tau**
- **Alzheimer's Disease (Important Mimic)**
- **FUS (Rare)**

Why This Matters

Symptoms alone do not tell us the cause.

- Different diseases can look **very similar clinically**
- The same symptoms can come from **different underlying biology**

How Diagnosis Works

There are currently no direct tests to confirm tau, TDP-43, or FUS during life

- Diagnosis is based on clinical evaluation, imaging, and pattern recognition
- Alzheimer's disease is the exception, with available biomarker testing

Treatment

There are **no disease-modifying treatments yet for Tau, TDP-43 or FUS**

Clinical trials are ongoing, with the goal of developing targeted treatments in the future

Is FTLD Genetic?

Some cases are inherited. The **majority of cases are not** and occur without family history

What This Means

FTLD reflects **biological changes in the brain**, not personality or choice.

Understanding the cause helps guide diagnosis, care, and planning.

Key Points

- FTLD is caused by different underlying proteins, not a single disease.
- These diseases affect brain networks, not just locations.
- Symptoms depend on which networks are disrupted (behavior, language, movement).
- The same disease can look different across people.
- Diagnosis focuses on

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