



Frontotemporal Dementia

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CLINICAL SYMPTOMS OF FRONTOTEMPORAL DEMENTIA

Frontotemporal dementia (FTD) is the second most common dementing illness in those under the age of 65. In general, it affects a younger population than Alzheimer's disease. Unlike Alzheimer's disease, FTD is more likely to start in the 50s-70s than in the 80s.

- FTD is an umbrella term, and your neurologist may give a more specific diagnosis based on each patient's history and symptoms. According to the most severe or the earliest symptoms, neurologists can sometimes give a more specific FTD diagnosis. These include:
 - **Behavioral variant FTD (bv-FTD)** – these patients have prominent, early changes in their behavior (such as being more impulsive or irritable), social interaction with others (kissing strangers, crude or explicit remarks), initiative to participate in hobbies (too much or too little), ability to control one's own actions (excessive eating, pacing), or decision making abilities. In the past, bv-FTD may have been called *Pick's disease* or *FTD*.
 - **Primary Progressive Aphasia (PPA)** – these patients have language abnormalities early in the disease course, although they can develop personality and behavior changes later. Depending on the type of language and speech problems in each patient, neurologists can further classify PPA into *semantic variant PPA* or *semantic dementia*, *agrammatic/non-fluent PPA* or *progressive non-fluent aphasia*, and *logopenic progressive aphasia*.
 - **Corticobasal syndrome (CBS)** – these patients have troubles with planning and execution of certain actions, and may have a hard time using familiar objects. Some of these patients may walk or move like they have Parkinson's disease, and some develop speech changes similar to PPA.
- Other symptoms may emerge in some patients later in the disease course, including symptoms of amyotrophic lateral sclerosis (ALS) or progressive supranuclear palsy (PSP).

CAUSES OF FRONTOTEMPORAL DEMENTIA

Scientists believe that the cause of a patient's dementia is related to the changes seen in his or her brain (the neuropathology). A number of unique neuropathologic changes have been identified in FTD, and many scientists at Emory and elsewhere are working on how to tell these changes apart by clinical examination, MRI, testing of blood or spinal fluid, or a combination of these tests.

When brains of patients with FTD are examined after death, we can divide them into certain groups:

- 10-15% of patients have changes similar to patients with **Alzheimer's disease**, even though their symptoms do not resemble those in Alzheimer's disease in older patients. A spinal fluid test for Alzheimer's disease may identify those in this group.
- Some patients show changes in a protein called TDP-43, collectively referred to as "**FTLD-TDP**".
- Some patients have changes associated with an abnormal form of the protein tau. Neurologists and pathologists refer to these cases collectively as "**FTLD-Tau**" or "**tauopathy**", although they may refine their diagnosis with more specific terms such as Pick's disease, corticobasal degeneration (CBD), progressive supranuclear palsy (PSP), etc.
- Other related diseases account for the remaining cases of FTD, including **FTLD-FUS**.

GENETICS/FAMILY HISTORY OF FRONTOTEMPORAL DEMENTIA

It is estimated that up to 40% of FTD patients have a family history of a progressive dementia or related disorder. Mutations in the following genes can lead to FTD: *PGRN* (for protein progranulin), *TARDBP* (for the protein TDP-43), *MAPT* (for the protein tau), and *VCP* (for the protein VCP).

DIAGNOSIS OF FRONTOTEMPORAL DEMENTIA

The main diagnosis of FTD and associated subtypes (bv-FTD, PPA, etc) is made by your neurologist after a complete evaluation which may include: history from you and your family members or friends; neurological examination; neuropsychological testing; brain imaging such as MRI; functional brain imaging such as a PET; blood tests for disorders that can cause behavior and/or language changes; lumbar puncture to look for causes of behavior and/or language changes not detectable in blood.

RESEARCH ON FRONTOTEMPORAL DEMENTIA AT EMORY

Emory investigators with an interest in FTD and related disorders include:

William Hu, MD, PhD	Nick Seyfried, DPhil	Allan Levey, MD, PhD	James Lah, MD, PhD
Jonathan Glass, MD	Jaffar Khan, MD	Stewart Factor, DO	Jorge Juncos, MD
Felicia Goldstein, PhD	Marla Gearing, PhD	Junmin Peng, PhD	

Current research projects related to FTD include:

- Natural history of FTD subtypes
- Development of a biomarker for FTLD-TDP and FTLD-Tau
- Cognitive decline in patients with amyotrophic lateral sclerosis (ALS)
- Cognitive decline in patients with progressive supranuclear palsy (PSP)
- Creation of a patient and caregiver support group for FTD

HOW TO TAKE PART

If you or someone you know has a diagnosis of FTD and would like to participate in research related to FTD at Emory, please contact the Emory Alzheimer's Disease Research Center at 404-728-6950

If you or someone you know would like a clinical evaluation, please contact 404-778-3444.