

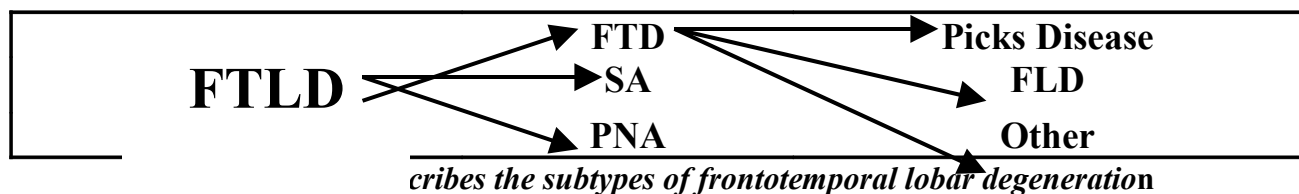
# FRONTOTEMPORAL DEMENTIA

## FREQUENCY

Frontotemporal lobar degeneration (FTLD) has received considerable public attention with claims that this disease is the second or third most common cause of intellectual loss (10-20%). Frontotemporal lobar degeneration dementia is a broad clinical term that describes patients with intellectual loss, psychiatric manifestations and evidence of frontal or temporal atrophy. Frontotemporal lobar degeneration has multiple pathological causes. Among them are frontal lobe degeneration (FTLD) and Pick's disease, as well as several other less commonly identified, pathological entities like semantic dementia and progressive non-fluent aphasia. Frontotemporal dementia (FTD) includes frontal lobe degeneration and Pick's disease. Many scientists speak as though frontotemporal dementia or frontotemporal lobar degeneration is a single disease that accounts for substantial numbers of dementia (10-15%); however, this clinical entity is a class of neurodegenerative disorder with multiple potential causes and pathologies (SEE BELOW). A very small number of FTD patients with Parkinsonism have genetic abnormalities linked to chromosome 17.

## TERMINOLOGY

Frontal lobe degeneration (FTD) is a common cause of dementia produced by degeneration of neurons in the frontal lobes. The clinical course averages 7 years and almost half the patients have a family member with a similar condition. Picks disease is bilateral (i.e., both sides) degeneration of a specific pair of lobes and the neuropathology is quite distinct. The clinical symptoms of Pick's disease depend on the location of damage. Semantic dementia and progressive nonfluent aphasia (PNA) are rare speech disorders. In conclusion, frontotemporal dementia usually applies to a group of patients who suffer from frontal lobe degeneration or Pick's disease.



## **SYMPTOMS**

The clinical symptoms of frontotemporal dementia differ from Alzheimer's disease because the FTD patient is a younger patient with more rapidly progressive dementia, and manifests more psychiatric symptomology. In comparison to Alzheimer's disease, the FTD patient has more apathy, disinhibition, impulsivity and decreased initiative. Patients may score well on the mini-mental status exam but perform poorly due to behavioral problems and other deficits. Brain imaging studies can document atrophy of the frontal or temporal lobes; however, substantial numbers of Alzheimer patients, alcoholic, and those with diffuse Lewy body disease also have frontal or temporal atrophy. No blood, urine, or spinal fluid is available to distinguish FTD from other clinical entities.

## **DIAGNOSIS**

Clinical history and neuropsychological testing are used to determine a possible diagnosis of FTD. Specialized brain-imaging tests such as SPECT or PET scans may prove useful. Alzheimer patients characteristically have parietal lobe abnormalities while FTD patients demonstrate frontal and temporal abnormalities. CT or MRI may show marked frontotemporal lobe degeneration.

## **THERAPY**

There is no present therapy for any type of frontotemporal dementia. Aricept, Exelon and other forms of treatment are not known to be helpful for patients with FTD. The protective role of estrogen, non-steroidal anti-inflammatory medications and antioxidant substances like vitamin E or ginkgo are not known to prevent the onset of these diseases. Psychiatric and behavioral symptom management is similar to treatment in Alzheimer's. Caregiver problems are similar to other types of dementia. Oftentimes, the apathy of FTD is confused with depression but this symptom is resistant to antidepressant therapy.

## **CAUSES**

Scientists attempt to understand the causes of dementia by postmortem studies on the brains of afflicted individuals who have clinical diagnoses. Many clinical research studies on living patients with FLTD or FTD lack pathological studies to confirm the diagnosis. The absence of brain tissues from well-characterized FTD patients minimizes the diagnostic accuracy and understanding of the disorder. Scientists presently have little understanding about the genetics of these disorders except for the very small group with linkage to chromosome 17.

Numerous, scientific problems occur when researchers lump multiple, pathological entities under a common clinical name. Frontotemporal dementia combines at least two pathological entities (frontal lobe degeneration and Pick's disease) without scientific proof that these diseases share a common cause or pathology. These questions will require large autopsy series that correlate clinical assessments of FTD to brain pathology. Family caregivers of patients with FTD should understand that this disease entity will undergo numerous changes and reconceptualization before clinicians agree upon a common terminology, pathology, and biology for these diseases.

## **A COMPARISON OF CLINICAL FEATURES FOR ALZHEIMER'S DISEASE AND FRONTOTEMPORAL DEMENTIA**

	<b>Alzheimer's Disease</b>	<b>Frontotemporal Dementia (FTD)</b>
<b>Age</b>	<b>Older <sup>a</sup></b>	<b>Younger <sup>a</sup></b>
<b>Onset</b>	<b>Insidious</b>	<b>Insidious</b>
<b>Progression</b>	<b>Yes</b>	<b>Yes</b>
<b>Cognitive Loss</b>		
<b>Fluency</b>	<b>++</b>	<b>++</b>
<b>Speech Output</b>	<b>++</b>	<b>+++</b>
<b>Naming</b>	<b>++</b>	<b>++</b>
<b>MMSE</b>	<b>++++ <sup>b</sup></b>	<b>++ <sup>b</sup></b>
<b>Calculations</b>	<b>+++</b>	<b>+</b>
<b>Construction</b>	<b>+++</b>	<b>+</b>
<b>Behavioral Abnormalities <sup>c</sup></b>		
<b>Apathy</b>	<b>+</b>	<b>++++</b>
<b>Disinhibition</b>	<b>+</b>	<b>++++</b>
<b>Depression</b>	<b>++</b>	<b>+</b>

**+ = minimal    ++ = mild    +++ = moderate    ++++ = severe**

**<sup>a</sup> = average 63 years for FTD and 75 years for AD**

**<sup>a</sup>Neary D, Snowden JS, Gustafson L, et al. Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. NEUROLOGY 1998;51:1546-1554.**

**<sup>b</sup> = average score 20.5 for FTD and 14.5 for AD**

**<sup>b</sup>Mendez MF, Cherrier M, Perryman KM, et al. Frontotemporal dementia versus Alzheimer's disease: Differential cognitive features. NEUROLOGY 1996;47:1189-1194**

**<sup>c</sup>Levy ML, Miller BL, Cummings JL, et al. Alzheimer's Disease and frontotemporal dementias: Behavioral Distinctions. ARCH NEUROL. 1996;53:687-690.**