Del Mar Caregiver Resource Center
A program of Health Projects Center

Fact Sheet

Frontotemporal Dementia

Definition

Frontotemporal Dementia (FTD) is a degenerative condition of the front (anterior) part of the brain. It differs from other causes of dementia such as Alzheimer's, Pick's and Creutzfeldt Jakob's diseases. The areas of the brain affected by FTD—the frontal and anterior temporal lobes—control reasoning, personality, movement, speech, social graces, language and some aspects of memory.

FTD is marked by dramatic changes in personality, behavior and some thought processes. Changes in personal and social conduct occur in early stages of the disease, including loss of inhibition, apathy, social withdrawal, hyperorality (mouthing of objects), and ritualistic compulsive behaviors. These symptoms may lead to misdiagnosis as a psychological or emotionally-based problem, or, in the elderly, be mistaken for withdrawal or eccentricity. FTD progresses to immobility and loss of speech and expression. Structural changes in the FTD patient's brain can be seen via scans or neuroimaging.

Facts

As many as seven million Americans may be afflicted with a form of dementia. Frontotemporal Dementia may account for 2-5 percent or 140,000 – 350,000 cases of dementia and for as many as 25 percent of pre-senile dementias.

FTD occurs predominantly after age 40 and usually before age 65, with equal incidence in men and women. In nearly half of the patients, a family history of dementia exists in a first degree relative (parent or sibling), suggesting a genetic component in these cases. Additionally, a form of dementia found in persons with motor neuron disease (amyotrophic lateral sclerosis, commonly known as "Lou Gehrig's Disease") may be associated with FTD.

Symptoms

Initial symptoms of FTD are primarily changes in personality and behavior. In addition to the symptoms described below, FTD patients often present two seemingly opposite behavioral profiles in the early and middle stages of the disease. Some individuals are overactive, restless, distractible and disinhibited. Others are apathetic, inert, aspontaneous and emotionally blunted. These differences in outward activity disappear in the late stages of the disease.

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Major symptoms of FTD are:

- Dramatic change in personal and social conduct. The individual may lack initiative, seem unconcerned, and neglect domestic, financial and occupational responsibilities.
- Loss of empathy toward others.
- Patients may show shallow affect (flat facial expression or lack of emotional response). Or they may be inappropriately jocular and sing, dance, clap or recite phrases repeatedly.

- Rigid and inflexible thinking and impaired judgment.
- Loss of insight into personal and social misconduct, such as small sexual or moral transgressions.
- Stereotyped (i.e., repetitive) or compulsive behavior. For example, the person with FTD may become compulsive about rituals of hygiene and dress while at the same time neglecting proper hygiene. They may echo what others say, wander restlessly over a fixed route, or adhere to a fixed daily schedule.
- Hypochondriasis, including bizarre somatic complaints.
- Excessive eating or gluttony, food fads (especially a craving for sweet foods) and even excessive alcohol consumption. (The tendency of FTD patients to consume alcohol often leads to a misdiagnosis of alcohol-related dementia.) The person may refuse to eat, however, due to a behavioral pattern called "negativism" or to inability to use motor skills needed for eating.
- Decreased motor skills in later stages.
- Change in sleep patterns, with prolonged sleepiness shown, especially in those that present more apathetic behaviors.

In late stage FTD symptoms include:

- A gradual reduction in speech, culminating in mutism.
- Hyperoral traits.
- Failure or inability to make motor responses to verbal commands.
- Akinesia (loss of muscle movement) and rigidity with death due to complications of immobility.

Differences Between FTD and Other Dementias

FTD differs markedly in several ways when compared to other dementias, especially Alzheimer's disease:

- FTD is characterized by cerebral atrophy in the frontal and anterior temporal lobes of the brain, while Alzheimer's affects the hippocampal, posterior temporal and parietal regions.
- The neurofibrillary tangles, senile plaques and Lewy bodies present in the brains of Alzheimer's and other dementia patients are absent. (Pick bodies are also usually absent.)
- Alzheimer's patients experience severe memory loss. While FTD patients exhibit memory disturbances, they remain oriented to time and place and recall information about the present and past.
- FTD patients, even in late stages of the disease, retain visuo-spatial orientation, and they negotiate and locate their surroundings accurately.
- Intellectual failure in FTD is distinctly different from that of Alzheimer's patients. Results of intelligence tests are normal in those with FTD until the point in the disease when disinterest results in lower scores.
- Life expectancy is slightly longer for FTD.

Testing and Diagnosis

FTD can be accurately diagnosed with brain scans or imaging. Computed tomography (CT scan) and magnetic resonance imaging (MRI) reveal cerebral atrophy in the frontotemporal regions. Degeneration of the corpus striatum, thalamus and other subcortical structures occurs. Functional brain imaging and single photon emission tomography may reveal dysfunction of the frontal lobes, decreased blood flow, and a selective reduced uptake of tracer in the anterior (front) cerebral hemispheres. Electroencephalography (EEG) remains normal, however, even in advanced stages. In autopsies, brain tissue changes include large neuronal cell loss with secondary spongiform change and astrocyctic gliosis.

Neuropsychological testing is useful to obtain a clinical assessment of the disease. Tests evaluate conduct, language, visuo-spatial abilities, memory, abstraction, planning and mental control, motor skills and intelligence. Tests might show:

- An economy of mental effort and unconcern. Responses may be impulsive and tasks readily abandoned, while other patients may be slow, inert and persistent.
- Conversation is not spontaneous; responses are brief and one does not elaborate. Some patients may make mechanical, repetitive remarks, echo words spoken by others, or repeat responses. Apathetic patients may be hypophonic (have a weak voice) and have an odd or halting speaking pattern. Overactive patients may be the opposite, with unconstrained speech. Failure to respond or inappropriate responses should not be assumed to be incomprehension, but rather a concreteness of thinking or inattention.
- Visuo-spatial skills remain normal except for those compromised by behavioral abnormalities.
- Memory problems do not occur except as a result of ineffective use of memory. Thought processes show impaired powers of abstraction, verbal response and design fluency. For example, in card and block sorting or picture arrangement, the FTD patient may abandon tasks, produce items eccentrically, not follow instructions, violate "rules," etc. Comprehension is normal, however.

Duration and Treatment

The length of FTD varies, with some patients declining rapidly over two to three years and others showing only minimal changes over a decade. Studies have shown persons with FTD to live with the disease an average of eight years, with a range from three years to 17 years.

No medications are known currently to treat or prevent FTD. Serotonin-boosting medications may alleviate some behaviors.

Credits

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Family Caregiver Alliance supports and assists caregivers of brain-impaired adults through education, research, services and advocacy.
FCA's information Clearinghouse covers current medical, social, public policy and caregiving issues related to brain impairments.

For residents of the greater San Francisco Bay Area, FCA provides direct family support services for caregivers of those with Alzheimer's disease, stroke, head injury, Parkinson's and other debilitating brain disorders that strike adults.

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