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Frontotemporal dementia

Frontotemporal dementia (FTD) is a rare form of dementia that is similar to Alzheimer disease, except that it tends to affect only certain areas of the brain.

Dementia may also be referred to as major neurocognitive disorder.

Causes

People with FTD have abnormal substances (called tangles, Pick bodies, Pick cells, and tau proteins) inside nerve cells in the damaged areas of the brain.

The exact cause of the abnormal substances is unknown. Many different abnormal genes have been found that can cause FTD. Some cases of FTD are passed down through families.

FTD is rare. It can occur in people as young as 20. But it usually begins between ages 40 and 60. The average age at which it begins is 54.

Symptoms

The disease gets worse slowly. Tissues in parts of the brain shrink over time. Symptoms such as behavior changes, speech difficulty, and problems with thinking occur slowly and get worse.

Early personality changes can help health care providers tell FTD apart from Alzheimer disease. (Memory loss is often the main, and earliest, symptom of Alzheimer disease.)

People with FTD tend to behave the wrong way in different social settings. The changes in behavior continue to get worse and are often one of the most disturbing symptoms of the disease. Some persons have more difficulty with decision-making, complex tasks, or language (trouble finding or understanding words or writing). There are variations of FTD seen with other nervous system problems such as:

- Amyotrophic lateral sclerosis (ALS/Lou Gehrig disease)
- Primary progressive aphasia
- Progressive supranuclear palsy

General symptoms include:

BEHAVIORAL CHANGES:

- Not able to keep a job
- Compulsive behaviors
- Impulsive or inappropriate behavior
- Inability to function or interact in social or personal situations
- Problems with personal hygiene
- Repetitive behavior
- Withdrawal from social interaction

EMOTIONAL CHANGES

- Abrupt mood changes
- Decreased interest in daily living activities
- Failure to recognize changes in behavior
- Failure to show emotional warmth, concern, empathy, sympathy
- Inappropriate mood
- Not caring about events or environment

LANGUAGE CHANGES

- Cannot speak (mutism)
- Decreased ability to read or write
- Difficulty finding a word
- Difficulty speaking or understanding speech (aphasia)
- Repeating anything spoken to them (echolalia)
- Shrinking vocabulary
- Weak, uncoordinated speech sounds

NERVOUS SYSTEM PROBLEMS

- Increased muscle tone (rigidity)
- Memory loss that gets worse
- Movement/coordination difficulties (apraxia)
- Weakness

OTHER PROBLEMS

- Urinary incontinence

Exams and Tests

Your provider will ask about the medical history and symptoms.

Tests may be ordered to check for other causes of dementia, including dementia due to metabolic causes. FTD is diagnosed based on symptoms and results of tests, including:

- Assessment of the mind and behavior (neuropsychological assessment)
- Brain MRI
- Electroencephalogram (EEG)
- Examination of the brain and nervous system (neurological exam)
- Examination of the fluid around the central nervous system (cerebrospinal fluid) after a lumbar puncture
- Head CT scan
- Tests of sensation, thinking and reasoning (cognitive function), and motor function
- Newer methods that test brain metabolism or protein deposits may better allow for more accurate diagnosis in the future
- Positron emission tomography (PET) scan of brain

Genetic tests can find mutations known to cause FTD and can support the diagnosis. A brain biopsy can also confirm the diagnosis, although it's usually never recommended.

Treatment

There is no specific treatment for FTD. Medicines may help manage mood swings.

Sometimes, people with FTD take the same medicines used to treat other types of dementia.

In some cases, stopping or changing medicines that worsen confusion or that are not needed can improve thinking and other mental functions. Such medicines include:

- Analgesics
- Anticholinergics
- Central nervous system depressants
- Cimetidine
- Lidocaine

It is important to treat any disorders that can cause confusion. These include:

- Anemia
- Decreased blood oxygen (hypoxia) level
- Heart failure
- High blood carbon dioxide level
- Infections
- Kidney failure
- Liver failure
- Nutritional disorders

- Thyroid disorders
- Mood disorders, such as depression

Medicines may be needed to control aggressive, dangerous, or agitated behaviors.

Behavior modification can help some people control unacceptable or dangerous behaviors. This consists of rewarding appropriate or positive behaviors and ignoring inappropriate behaviors (when it is safe to do so).

Talk therapy (psychotherapy) does not always work. This is because it can cause further confusion or disorientation.

Reality orientation, which reinforces environmental and other cues, may help reduce disorientation.

Depending on the symptoms and severity of the disease, monitoring and help with personal hygiene and self-care may be needed. Eventually, there may be a need for 24-hour care and monitoring at home or in a special facility. Family counseling can help the person cope with the changes needed for home care.

Care may include:

- Adult protective services
- Community resources
- Homemakers
- Visiting nurses or aides
- Volunteer services

People with FTD and their family may need to seek legal advice early in the course of the disorder. Advance care directive, power of attorney, and other legal actions can make it easier to make decisions regarding the care of the person with FTD.

Support Groups

You can ease the stress of FTD by joining a support group. Sharing with others who have common experiences and problems can help you not feel alone. More information and support for people with FTD and their families can be found at:

The Association for Frontotemporal Degeneration -- www.theaftd.org/get-involved/in-your-region/
[<https://www.theaftd.org/get-involved/in-your-region/>]

Outlook (Prognosis)

The disorder steadily becomes worse. The person becomes totally disabled early in the course of the disease.

FTD commonly causes death within 8 to 10 years, usually from infection, or sometimes because other body systems fail.

When to Contact a Medical Professional

Contact your provider or go to the emergency room if mental function gets worse.

Prevention

There is no known prevention.

Alternative Names

Semantic dementia; Dementia - semantic; Frontotemporal dementia; FTD; Arnold Pick disease; Pick disease; 3R tauopathy; Frontotemporal lobar degeneration (FTLD)

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