

Introduction

Alzheimer's disease is the most common of a large group of disorders known as "dementias." It is an irreversible disease of the brain in which the progressive degeneration of brain cells causes thinking ability and memory to deteriorate. Alzheimer's disease also affects behaviour, mood and emotions, and the ability to perform daily living activities.

Other forms of dementia resemble Alzheimer's disease in that they also involve a progressive degeneration of brain cells that is currently irreversible. They include the dementia associated with vascular dementia (the second most common dementia after Alzheimer's disease), frontotemporal dementia, Creutzfeldt-Jakob disease, Lewy body dementia, Huntington disease, and Parkinson's disease.

Sometimes a person may have different symptoms in the early stages of the disease, such as memory loss, behaviour changes, or difficulties with speech and movement. These symptoms may suggest a form of dementia other than Alzheimer's disease. Frontotemporal dementia (FTD) is one of these dementias. In any event a person should always seek a thorough medical assessment if any of these symptoms are present.

Regardless of the type of dementia, individuals are encouraged to obtain information and support from the Alzheimer Society.

What is frontotemporal dementia?

Unlike Alzheimer's disease, which generally affects most areas of the brain, frontotemporal dementia is an umbrella term for a group of disorders that primarily affect the frontal and temporal lobes of the brain - the areas generally associated with personality, behaviour, emotions, language, speech, abstract thinking and movement. In most cases of FTD, brain cells in these areas shrink or die. In other cases, the brain cells in these areas get larger, containing round, silver "Pick's bodies". The diagnosis is then Pick's disease or behavioural variant Frontotemporal Dementia (bvFTD), the most common frontotemporal disorder.

Frontotemporal dementia is a progressive disease and according to researchers, approximately 2-5% of all dementia cases are frontotemporal dementia. However, it likely accounts for 20% or more of dementia in individuals under the age of 65.

Other names often used for frontotemporal dementia, in addition to Pick's disease, include: frontal lobe dementia, frontotemporal lobar degeneration and Pick's complex, Tau disease, Semantic dementia, non-fluent primary progressive aphasia, corticobasal degeneration and progressive supranuclear palsy. In general, they all eventually lead to similar physical signs or symptoms associated with FTD. FTD involves changes in personality, behaviour, and judgment. People with this dementia can act inappropriately around other people, resulting in embarrassing social situations. In the early stage of FTD, behaviour changes and/or problems

with speech (language) can appear separately. As the disease progresses, these two areas will overlap, language problems will progress and affect the ability of the person to communicate.

Over time, the language and/or movement problems may occur, and the person may need more care and supervision¹. Unlike Alzheimer's disease, a person with frontotemporal dementia often remains oriented with respect to time and has preserved memory in the early stages. In the later stages of the disease, general symptoms of dementia arise, i.e. confusion and forgetfulness, as in other dementias.

How does frontotemporal dementia affect the person?

Since the frontal and temporal areas of the brain can be affected, symptoms often affect personality, behaviour, emotions, language, speech, abstract thinking and movement.

- **Changes in behaviour** may include becoming either withdrawn or disinhibited (e.g., losing the ability to restrain one's behaviour and actions). The person may show reduced initiative and lose interest in personal hygiene, become easily distracted or repeat the same action over and over again. Overeating or compulsively putting objects in the mouth may occur. Sometimes incontinence is an early symptom of the disease. People with FTD may become indifferent toward other people or may experience abrupt and frequent mood changes or excessive emotions.
- **Problems with language** can range from trouble finding the right word or reduction of speech to total loss, i.e. becoming mute. Echoing what has been said by others and stuttering are common symptoms. The person may have difficulty sustaining a train of thought or maintaining a conversation for any length of time. Writing and reading are also affected.
- **Problems with movement.** People with FTD may experience decreased facial expression, slowness of movements, rigidity and postural instability. Shaking, difficulty walking, frequent falls, and poor coordination can be common. Motor skills are lost and swallowing difficulties may occur.

How is frontotemporal dementia assessed?

No single test can diagnose frontotemporal dementia. Doctors diagnose the disease through a process of identifying characteristic features of the disease and ruling out other possible causes.

The diagnosis of frontotemporal dementia generally involves:

- Medical history, brain imaging (such as MRI (magnetic resonance imaging), PET (positron emission tomography), and SPECT (single photon emission computed tomography) and detailed neurological examination
- Physical and neuropsychological examination to assess language, behaviour, memory, executive, and visual-spatial functions

¹ *Frontotemporal disorders: Information for patients, families and caregivers*, National Institute on Aging, September 2010.

What are the risk factors for frontotemporal dementia?

Frontotemporal dementia tends to occur at a younger age than Alzheimer's disease and can affect both men and women. The average length of the disease is 2-10 years. Little is known about the cause of frontotemporal dementia and risk factors have yet to be identified. Recent research has shown genetic mutations underlying some cases of FTD. However, more than half the people who develop frontotemporal dementia have no family history of dementia.

Is there treatment?

At present, there is no known cure and no effective way to slow its progression. Cholinesterase inhibitors used as treatment for Alzheimer's disease do not work for the treatment of FTD as different areas of the brain are affected. Treatment currently focuses on managing symptoms.

New behavioural therapeutic strategies are also helping people living with the disease. Therapeutic techniques like physical activity and music are being used as viable and useful treatments. Research shows that the quality of life of people with dementia, and also their caregivers, is significantly improved by activities that emphasize their strengths and abilities. By understanding the person's personality, life experiences, support systems and ways of coping, a person-centred approach to care can be created that preserves and improves quality of life.

For more information:

Visit the Alzheimer Society's website at www.alzheimer.ca or contact your local Alzheimer Society.

More information can be obtained from the following:

- The Association for Frontotemporal Degeneration: www.theaftd.org
- Frontotemporal disorders: Information for patients, families and caregivers, National Institute on Aging. http://www.nia.nih.gov/sites/default/files/ftd_2014_update-final-lowres.pdf
- Pick's Disease Support Group: www.pdsg.org.uk
- Mayo Clinic: www.mayoclinic.com/health/frontotemporal-dementia/DS00874
- UCSF Memory and Aging Center: <http://memory.ucsf.edu/ftd/>

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Alzheimer *Society*

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