

Related Dementia - Frontotemporal Dementia

Introduction

Dementia is a syndrome consisting of a number of symptoms that include loss of memory, judgment and reasoning, and changes in mood, behaviour and communication abilities. These symptoms may affect a person's ability to function at work, in social relationships or in day-to-day activities. Sometimes symptoms of dementia can be caused by conditions that may be treatable, such as depression, thyroid disease, infections or drug interactions. However, if the symptoms are not treatable and progress over time, they may be due to damage to the nerve cells in the brain.

Alzheimer's disease is the most common form of dementia. It is a disease of the brain, characterized by deterioration of thinking ability and of memory, caused by the progressive death of brain cells. Alzheimer's disease accounts for approximately 64% of all dementias in Canada. The features of Alzheimer's disease include a gradual onset and continuing decline of memory, as well as changes in judgment or reasoning, and ability to perform familiar tasks. Other dementias include Vascular Dementia, Lewy body Dementia, Frontotemporal Dementia, and Creutzfeldt-Jakob Disease.

Sometimes a person may have symptoms such as sudden onset of memory loss, behaviour changes, or difficulties with speech and movement. These symptoms may suggest a dementia other than Alzheimer's disease. Frontotemporal Dementia is one of these dementias. A person should 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 rare disorders that primarily affect the frontal and temporal lobes of the brain - the areas generally associated with personality and behaviour. In some cases, brain cells in these areas can shrink or die. In other cases, the brain cells in these areas get larger, containing round, silver "Pick's bodies." The term "Pick's Disease" is used to describe this subtype of Frontotemporal Dementia that has these specific abnormalities. In both situations, the changes in the brain affect the person's ability to function. Researchers estimate that approximately 2% of all dementia cases are Frontotemporal Dementia.

Other names often used for Frontotemporal Dementia include: Pick's Disease, Semantic Dementia, Frontal Lobe Dementia, Primary Progressive Aphasia, Corticobasal Degeneration and Pick's Complex.

How does Frontotemporal Dementia affect the person?

Since the frontal and temporal areas of the brain can be affected in different ways, early symptoms often affect either **behaviour** and/or **speech (language)**.

- **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 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.

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- **Problems with speech (language)** can range from 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.

In the early stage of Frontotemporal Dementia, behaviour changes or problems with speech (language) can appear separately. As the disease progresses, these two areas will overlap. Unlike Alzheimer's disease, a person with Frontotemporal Dementia often remains oriented 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. Motor skills are lost and swallowing difficulties 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. Brain imaging (such as with an MRI) can also be helpful in making a diagnosis of Frontotemporal Dementia.

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. While most cases are not inherited, there is an autosomal dominant gene (chromosome 17) which can be passed from generation to generation. This type of Frontotemporal Dementia is extremely rare.

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, usually do not work for the treatment of Frontotemporal Dementia, as different areas of the brain are affected. Treatment currently focuses on managing symptoms.

For more information:

Visit the Alzheimer Society of Toronto's website www.alzheimertoronto.org or call us 416-322-6560.

More information can be obtained from the following:

- Pick's Disease Support Group: www.pdsg.org.uk/
- Mayo Clinic: <http://www.mayoclinic.com/health/frontotemporal-dementia/DS00874>

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