

FRONTOTEMPORAL DEMENTIA

This Help Sheet describes a type of dementia known as frontotemporal dementia, which has different forms including behavioural-variant frontotemporal dementia, progressive non-fluent aphasia and semantic dementia.

What is frontotemporal dementia?

Frontotemporal dementia (FTD) is the name given to dementia when it is due to progressive damage to the frontal and/or temporal lobes of the brain.

The right and left frontal lobes at the front of the brain are involved in mood, social behaviour, attention, judgement, planning and self-control. Damage can lead to reduced intellectual abilities and changes in personality, emotion and behaviour.

The right and left temporal lobes at the two sides of the brain are involved in processing what we hear and understanding what we hear and see. Damage may lead to difficulty recognising objects or understanding or expressing language.

FTD is sometimes called frontotemporal lobar degeneration (FTLD). It was first described 100 years ago by Arnold Pick and was previously referred to as Pick's disease.

The symptoms of FTD depend on which areas of the brain are damaged. In contrast to Alzheimer's disease, memory often remains unaffected in FTD, especially in the early stages. When the frontal lobes are affected first, the main changes are in personality and behaviour, and this is called behavioural-variant FTD. When the temporal lobes are affected first, there is a loss of language skills. There are two types of FTD where language is impaired – progressive non-fluent aphasia and semantic dementia.

Behavioural-variant frontotemporal dementia

In the frontal or behavioural variant of FTD, there are changes in the person's behaviour, habits, personality and/or emotional responses. Symptoms vary from person to person depending on which areas of the frontal lobes are damaged. Some people with behavioural-variant FTD become very apathetic, while others become disinhibited.

Common symptoms can include:

- Fixed mood and behaviour, appearing selfish and unable to adapt to new situations
- Loss of empathy, emotional warmth and emotional responses

- Apathy or lack of motivation, abandoning hobbies or avoiding social contact
- Loss of normal inhibitions, talking to strangers or exhibiting embarrassing behaviour
- Difficulty in reasoning, judgement, organisation and planning
- Distractibility and impulsiveness
- Changes in eating patterns, craving sweet foods, overeating or unusual food preferences
- A decline in self-care and personal hygiene
- Lack of insight

Reduced ability to perform daily activities is an early feature. As the disease progresses, the person may become obsessive, or repeat patterns of movement or behaviours.

Semantic dementia

In the temporal lobe form of FTD, the initial symptom is usually a decline in language abilities. In semantic dementia, the ability to assign meaning to words is gradually lost. Reading, spelling, comprehension and expression are usually affected.

Semantic knowledge or memory refers to the meanings of words, objects and concepts. People with semantic dementia experience impaired understanding of single words, especially uncommon words. They experience difficulty finding words and people's names. Grammar and speech production remain intact, so speech still sounds fluent but may seem empty in content.

If the right temporal lobe is involved the person may have problems recognising previously familiar people. Many people with semantic dementia retain non-language abilities until very late in the disease. In later stages, the disease spreads to the frontal lobes and changes in emotional responses, empathy and food preferences are common.

Progressive non-fluent aphasia

Progressive non-fluent aphasia (PNFA) is the least common form of FTD and tends to have a later onset. The ability to speak fluently is gradually lost. People with PNFA have difficulty communicating due to slow

and difficult production of words, distortion of speech and a tendency to produce the wrong word.

Some people with PNFA have slurring of speech, whereas others are articulate but say the wrong word. Understanding of word meaning is preserved and following conversations is difficult. Using the telephone and communicating with groups of people are particularly difficult.

Spelling is often impaired and some people develop difficulties reading. Subtle deficits in problem solving, mental flexibility and decision making are often present. In later stages, the disease spreads to the frontal lobes so that behavioural changes occur, especially changes in emotional responses and empathy.

Overlap between frontotemporal dementia and movement disorders

Most people with FTD do not experience significant movement or motor problems. However, motor symptoms can develop in the later stages of the illness and FTD can co-occur with other conditions characterised by motor symptoms. These include motor neuron disease, corticobasal syndrome, progressive supranuclear palsy and FTDP-17 (FTD with parkinsonism linked to chromosome 17).

Who gets frontotemporal dementia?

FTD can affect anybody. It typically affects people at a younger age than Alzheimer's disease, with symptoms beginning in the 50s or 60s, and sometimes younger. Almost a third of people with FTD have a family history of dementia. However, only about 10-15% of cases have familial FTD, in which a gene mutation is passed on that causes the disease. The genetic basis of FTD is not fully understood and is actively being researched.

How is frontotemporal dementia diagnosed?

It is important that someone with suspected FTD is assessed by a neurologist, geriatrician or psychiatrist specialising in dementia to establish the diagnosis. A typical assessment would include a detailed medical history, a physical examination, blood and urine tests, a psychiatric assessment, a neuropsychological assessment and brain imaging.

The presence of changes in the frontal and temporal lobes on MRI scans is critical for an accurate diagnosis. Neuropsychological tests to identify specific problems in areas such as comprehension and problem solving also help establish the diagnosis.

What causes frontotemporal dementia?

FTD is caused by brain disease, but why some people get the disease is unknown (except in familial FTD

which is caused by a genetic mutation). People with FTD can have one of a number of different underlying changes in brain cells. These various cellular changes generally can not be observed during life, but only with a brain autopsy to identify the changes under a microscope.

Cellular studies of the brain have shown that there are two types of protein which accumulate in brain cells in FTD – tau and TDP-43. These protein accumulations damage and kill brain cells in the frontal and/or temporal lobes. As the disease progresses, these regions show shrinkage detectable on MRI scans.

How does frontotemporal dementia progress?

Although people with FTD may be assessed as having one of the three subtypes above, the disease will progress and symptoms of 2 or 3 subtypes are likely to occur. FTD causes progressive and irreversible decline in a person's abilities over a number of years, and is a terminal disease.

Is there treatment available?

There is not yet a cure for FTD, or any disease-modifying treatment. However, various therapies can help with some of the symptoms. Secondary symptoms, such as depression, may be helped by medication.

Knowing more about FTD and why the person is behaving as they are can help people to cope with the disease. Family members and carers can, with support, develop coping strategies to work around problems rather than trying to change the behaviour of the person with FTD.

Psychological therapies are important to help manage abnormal behaviour. Speech therapy is of benefit to people with PNFA, particularly in helping to develop alternative communication methods. The input of an occupational therapist can help improve everyday functioning at home.

More information about FTD is available from the websites of the Frontotemporal Dementia Research Group (Frontier) at neura.edu.au, the Association for Frontotemporal Degeneration at theftd.org and the FTD Toolkit at ecdc.org.au/ftd-toolkit.htm

FURTHER INFORMATION

Alzheimer's Australia offers support, information, education and counselling. Contact the National Dementia Helpline on 1800 100 500, or visit our website at fightdementia.org.au



Interpreter

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