FRONTOTEMPORAL DEMENTIA

Dementia (also known as Neurocognitive Disorder) causes gradual deterioration in attention, decision-making, memory and learning, language, perception and /or social behaviour.

What is frontotemporal dementia?

Frontotemporal dementia (FTD) is an umbrella term for a group of disorders affecting the frontal and temporal lobes of the brain and is the cause of at least 5% of all dementia cases, possibly more since it is difficult to diagnose. The frontal and temporal regions of the brain can be affected by other forms of dementia. For example, Alzheimer's disease can start in the frontal region and poor blood supply to these lobes can occur in vascular dementia. However, generally there are three recognised forms of frontotemporal dementia.

1. **Behavioural variant frontotemporal dementia**
   In this condition there are early changes in behaviour, personality and executive function, that is, poor reasoning, judgement and decision- making. The behaviour and personality changes may include apathy, disinhibition (e.g., tactless, socially inappropriate behaviour), loss of empathy and social interest. The person may develop repeated, pointless movements or sayings. Eating patterns change and the person shows a tendency to always put something in their mouth (hyperorality). The person is not usually aware of problems and may make disastrous personal or financial decisions due to poor judgement and loss of inhibitions.

2. **Language variants of frontotemporal dementia**
   There are two sorts of Frontotemporal dementia that initially affect language, called primary progressive aphasia.

   (a) *In the semantic variant* the first symptoms are a decline in language skills so that the person has trouble producing words, naming things and structuring sentences. People may struggle to find and understand and pronounce words, especially the ones they do not use very often. The meaning of words can be lost, but grammar remains intact.

   (b) *Non-fluent variation* of primary progressive aphasia: In this condition, speech is slow, laboured and halting and words may be left out and grammar misused. People might have trouble under standing complex sentences. People can retain their writing skills for a long time, but spelling and reading can be impaired.

Overlap with movement disorders

Perhaps 40% of all people with Frontotemporal dementia develop mild motor neuron symptoms such as weakness, stiffness, difficulties with speech and swallowing and changes in emotional expression. About 20% get Parkinson's-like symptoms. Others experience eye movement problems, instability standing and falls.

Causes

Why these changes occur in the brain is unknown.

Who gets it?

Frontotemporal dementia is a common cause of dementia in people younger than 65, only about 25% of cases occur in people over 65 (DSM V). About 40% of people with Frontotemporal dementia have a family history of early onset dementia and some specific genes have been identified.

Diagnosis

History: The diagnosis is made in the usual manner by the doctor asking the person and other people who know him/her about the onset and course of symptoms. It is important to find out if there is a family history of similar problems. Because it is often difficult to tell Frontotemporal dementia from common psychiatric disorders, a GP might ask for a psychiatrist's opinion.

Laboratory and radiology: This will test for other causes of impairment. A CT or MRI may show specific changes confirming Frontotemporal dementia or another form of dementia.

Neuropsychological testing: This can indicate abnormalities in the frontal lobe or elsewhere. If there are mainly memory and spatial perception defects and social appropriateness is maintained then the diagnosis is more likely to be Alzheimer's disease, affecting the frontotemporal regions.

How does Frontotemporal dementia progress?

Frontotemporal dementia is progressive, spreading to the rest of the brain eventually looking like other forms of dementia. The rate of decline tends to be faster than the other dementias with a shorter survival time after diagnosis.

Treatment

There is no specific treatment for Frontotemporal dementia except for addressing the symptoms. Treatment with cholinesterase inhibitors is not beneficial in any of these conditions and may worsen behavioural symptoms. Memantine makes no difference to the progression of Frontotemporal dementia symptoms. Certain antidepressants, selective serotonin re-uptake inhibitors (SSRIs) can reduce the severity of compulsions, aggression, abnormal eating, impulsiveness, and agitation. Sometime low doses of atypical antipsychotics help some of the behaviour abnormalities but need to be used cautiously.