Dementia is a term used to describe a loss of mental abilities. The commonest cause is Alzheimer's disease. This is associated with nerve cell loss and atrophy ('shrinkage') of the temporal and parietal lobes of the brain. In the rarer **fronto-temporal dementias (FTD**), the atrophy is in the frontal and/or temporal lobes. These areas of the brain are involved particularly with behaviour, personality and language, so it is these functions rather than memory and orientation which are most affected initially.

One form of FTD is **primary progressive aphasia (PPA).** This describes patients with deteriorating language skills, but with other brain functions relatively intact, at least until later stages of the illness. **Semantic dementia** causes problems with the meaning and content of language. The **behavioural variant** of FTD presents with personality and behavior change. **Pick's disease** is a specific form of FTD, associated with abnormal cells seen on microscopic examination of brain tissue after death.

What symptoms does FTD cause?

The early symptoms of FTD are often confused with depression or Alzheimer's disease. FTD has a gradual onset and slow progression. Memory and perceptual skills remain relatively preserved until late in the disease. Patients usually look physically well, but urinary problems may sometimes develop early and change in food preferences (typically a liking for sweets) may lead to significant weight change.

Other characteristic features are:-

- **Behavioural disturbance**. This may be the initial symptom and presents as a deterioration in social conduct, disinhibition and neglect of personal hygiene and daily routine. There may be childlike obsessions or repetitive rituals.

- **Personality and mood change**. There may be uncharacteristic and insensitive jokiness or rudeness, increasing selfishness, apathy, remoteness and lack of emotional warmth. Early loss of insight is typical.

- **Lack of initiative, inattention, poor planning skills and poor judgment**. Thinking becomes more rigid and inflexible.

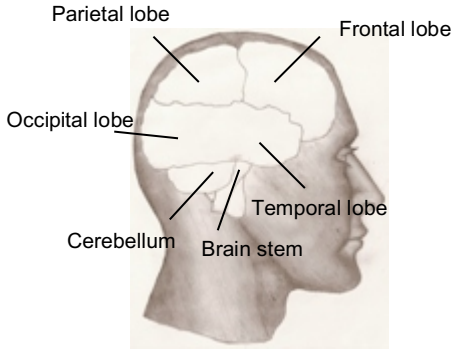
- **Conversation is affected**, with loss of fluency, tendency to use the same phrase or sentence over again (echoing), or circumlocution with many words but little content.

How common is FTD?

FTD presents at an earlier age than Alzheimer's disease and typically affects people in their 50s and 60s. About 5% of patients with dementia of all ages will have FTD, but up to 20% or more of those aged less than 65 years. The course is variable, but the average rate of decline in FTD may be faster than in Alzheimer's disease.

Who gets FTD and why?

In most cases of FTD, the cause is unknown. However, there is a family history in up to half of all cases and it may then be possible to identify the faulty gene, after counselling from a geneticist.



How is FTD diagnosed?

The diagnosis of FTD is based on the clinical presentation, supported by results of specialised neuropsychological tests. Brain scans may demonstrate localised wasting or atrophy of the frontal and/or temporal lobes. However, scans are not diagnostic and may be normal in the early stages of the illness. In people with a family history, it may be possible to identify the faulty gene causing the disease. A few people may also have features of motor neurone disease.

Usually a definite diagnosis of FTD can only be made after death by detecting characteristic appearances of brain tissue under the microscope. Some patients have specific abnormalities ('Pick bodies') within swollen nerve cells ('Pick cells'). Others have mainly loss of brain cells in the frontal and temporal lobes.

What treatment is available?

Unfortunately there is no treatment to cure or to stop progression of the illness. As in other types of dementia, management centres on good medical, psychological and social care and support and education for the patient and family. Certain drugs sometimes may help with behavioural problems, but must be used under careful supervision. The anticholinesterase drugs used in Alzheimer's disease are not beneficial in FTD and may make symptoms worse.

Where can you get more information about FTD?

The Rare Dementia Support Group is based in London and has information about FTD for patients and families available at:
www.raredementiasupport.org/ftd

The Alzheimer's Society
http://alzheimers.org.uk/
has information on all aspects of dementia.

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Fronto-temporal dementia

CARDIFF MEMORY TEAM

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