

What is frontotemporal dementia (FTD)?

Factsheet 404LP
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Frontotemporal dementia (FTD) is one of the less common types of dementia. The term covers a wide range of different conditions. It is sometimes called Pick's disease or frontal lobe dementia.

This factsheet explains what FTD is, its symptoms, and who gets it. It also describes how it is diagnosed and the treatment and support that is available.

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What is frontotemporal dementia (FTD)?

The word 'frontotemporal' refers to the lobes of the brain that are damaged in this type of dementia. The frontal lobes of the brain, found behind the forehead, deal with behaviour, problem-solving, planning and the control of emotions. An area of usually the left frontal lobe also controls speech.

The temporal lobes – on either side of the brain – have several roles. The left temporal lobe usually deals with the meaning of words and the names of objects. The right temporal lobe is usually involved in recognising faces and familiar objects. For more see factsheet 456, **Dementia and the brain**.

Frontotemporal dementia occurs when nerve cells in the frontal and/or temporal lobes of the brain die, and the pathways that connect the lobes change. Some of the chemical messengers that transmit signals between nerve cells are also lost. Over time, as more and more nerve cells die, the brain tissue in the frontal and temporal lobes shrinks.

When the frontal and/or temporal lobes are damaged in this way, this causes the symptoms of FTD. These include changes in personality and behaviour, and difficulties with language. These symptoms are different from the memory loss often associated with more common types of dementia, such as Alzheimer's disease.

As FTD is a less common form of dementia, many people (including some health professionals) may not have heard of it.

Frontotemporal dementia and younger people

Frontotemporal dementia is much less common than other forms of dementia, such as Alzheimer's disease or vascular dementia. However, it is a significant cause of dementia in younger people – that is, those under the age of 65. Frontotemporal dementia is probably the third most common cause of dementia in this age group and some studies even place it second most common. It affects men and women roughly equally.

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Frontotemporal dementia is most often diagnosed between the ages of 45 and 65. However, it can also affect people younger or older than this, and it is probably under-recognised in older people. Even so, this 'peak age' for FTD (the age at which it is most often diagnosed) is much younger than the age at which people are most often diagnosed with the more common types of dementia, such as Alzheimer's disease.

Being diagnosed at a younger age is likely to present someone with a different set of challenges. They may still be working, have financial commitments or dependent children, and want different services and support. For more information about these issues see factsheet 440, **What is young-onset dementia?**

Symptoms

The symptoms of FTD dementia vary depending on which areas of the frontal and temporal lobes are damaged. A person may have one of three main types of FTD:

- behavioural variant FTD
- progressive non-fluent aphasia
- semantic dementia.

As with most forms of dementia, the symptoms can be very subtle at first, but they slowly get worse as the disease progresses.

Behavioural variant FTD

This is the most common type of FTD. Two thirds of people with FTD are diagnosed with this type. During the early stages, changes are seen in the person's personality and behaviour.

A person with behavioural variant FTD may:

- lose their inhibitions – behave in socially inappropriate ways and act in an impulsive or rash manner. This could include making tactless or inappropriate comments about someone's appearance

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- lose interest in people and things (apathy) – lose motivation, but (unlike someone with depression) they are not sad
- lose sympathy or empathy – become less responsive to the needs of others and show less social interest or personal warmth. They may also show reduced humour or laugh at other people's misfortunes. This can make the person appear selfish and unfeeling
- show repetitive, compulsive or ritualised behaviours – this can include repeated use of phrases or gestures, hoarding and obsessions with timekeeping. It may also include new interests, such as music or spirituality
- crave sweet, fatty foods or carbohydrates and forget table etiquette. They may also no longer know when to stop eating, drinking alcohol or smoking.

It is common for a person with behavioural variant FTD to struggle with planning, organising and making decisions. These difficulties may first appear at work or with managing finances.

In contrast to those with Alzheimer's disease, people in the early stages of behavioural variant FTD tend not to have problems with day-to-day memory or with visuospatial skills (judging relationships and distances between objects). Someone with FTD may go walking without obvious purpose but, unlike a person with Alzheimer's, will often find their way home without getting lost.

Recent research shows that FTD can also affect the sensitivity of people with dementia to physical or environmental stimulation such as temperature, sounds and even pain.

It is unusual for a person with behavioural variant FTD to be aware of the extent of their problems. Even early on, people generally lack control over their behaviour or insight into what is happening to them. Their symptoms are more often noticed by the people close to them.

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Language variants of FTD

In the other two types of FTD, the early symptoms are difficulties with language that progressively get worse. These difficulties become apparent slowly, often over two or more years.

In progressive non-fluent aphasia, these problems are with speech – ‘aphasia’ means loss of language. Common early symptoms may include:

- slow, hesitant speech which may seem difficult to produce – a person may stutter before they can get the right word out, or may mispronounce it when they do
- errors in grammar – a person may have ‘telegraphic speech’, leaving out small link words such as ‘to’, ‘from’ or ‘the’
- impaired understanding of complex sentences, but not single words.

In semantic dementia, speech is fluent but people begin to lose their vocabulary and understanding of what objects are. Common early symptoms may include:

- asking the meaning of familiar words (eg, ‘What is “bread”?’)
- trouble finding the right word, leading to descriptions instead (eg ‘the thing for opening tins’), or use of less precise words (eg ‘animal’ instead of ‘cat’)
- difficulty recognising familiar people or common objects.

In the early stages of both language forms of FTD, other mental abilities (such as memory, visuospatial skills, planning and organising) tend to be unaffected.

Later stages

The rate at which FTD progresses varies greatly, with life expectancy ranging from less than two years to 10 years or more. Research shows that on average, people live for about six to eight years after the start of symptoms but this varies widely.

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As FTD progresses, the differences between the three types become much less obvious. People with the behavioural variant tend to develop language problems as their condition progresses. They may eventually lose all speech, like a person with one of the language variants.

Similarly, over several years a person with a language variant of FTD (especially semantic dementia) will tend to develop the behavioural problems typical of behavioural variant FTD.

In the later stages of all types of FTD, more of the brain becomes damaged. As a result, the symptoms are often similar to those of the later stages of Alzheimer's disease. The person may become less interested in people and things and have limited communication. They may become restless or agitated, or behave aggressively. At this late stage, they may no longer recognise friends and family, and are likely to need full-time care to meet their needs.

Overlapping motor disorders

About 10–20 per cent of people with FTD also develop a motor disorder, either before or after the start of dementia. A motor disorder is one that causes difficulties with movement. These motor disorders, which are generally uncommon but more likely in people with this form of dementia, are:

- motor neurone disease
- progressive supranuclear palsy
- corticobasal degeneration.

The symptoms of these three conditions are similar and can include twitching, stiffness, slow movements and loss of balance or co-ordination. In the later stages, they can often cause difficulties with swallowing. Progressive supranuclear palsy and corticobasal degeneration share some symptoms with Parkinson's disease and are sometimes called 'atypical parkinsonism'. For more information see factsheet 442, **Rarer causes of dementia**.

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These motor disorders are all degenerative diseases of the nervous system, meaning that they will get worse over time. If a person has both FTD and motor neurone disease, they can deteriorate more quickly than someone with FTD alone. On average, a person with both conditions will live for two or three years after diagnosis.

Causes

We don't know exactly what causes FTD. Experts think that the disease is due to a mixture of genetic, medical and lifestyle factors. Even allowing for its under-recognition in older people, FTD does not show the very strong link with ageing seen for more common dementias such as Alzheimer's disease or vascular dementia.

Autopsy studies show that the death of nerve cells in the frontal and temporal lobes is linked to clumps of abnormal proteins inside the cells, including proteins called tau and TDP-43. The tau protein may take the form of Pick bodies, which gave FTD its original name of Pick's disease – after Dr Arnold Pick who first studied the dementia.

Frontotemporal dementia is much more likely to run in families than the more common forms of dementia are. About one third of people with the condition have some family history of dementia.

About 10–15 per cent of people with FTD have a very strong family history of the condition, with several close relatives in different generations affected. This pattern is most common in the behavioural type of FTD and least common in semantic dementia. Typically in these cases, FTD is inherited from a parent as a defect (mutation) in one of three genes: MAPT, GRN or C9ORF72.

Each of the children or siblings of someone with a mutation that is known to cause FTD has a 50 per cent chance of carrying the same mutation. Families with a known mutation should be offered a referral to a specialist genetics service for counselling. For more about genetic testing see factsheet 405, **Genetics of dementia**.

Diagnosis

Frontotemporal dementia can be hard to diagnose, because it is relatively uncommon and does not initially cause memory problems. The person with FTD may lack insight into their behaviour, and so they may not want to seek professional help. Doctors may also not suspect dementia in what is often a middle-aged person.

Frontotemporal dementia may be misdiagnosed as atypical Alzheimer's disease (a form of Alzheimer's disease in which people don't have early memory loss). The behavioural symptoms may easily be mistaken for depression, schizophrenia or obsessive-compulsive disorder. Problems with language or movement may be misdiagnosed as stroke. Blood tests and a thorough physical examination are important to rule out other possible causes of symptoms. A specialist may suspect a diagnosis of FTD after questioning the affected person and someone who knows them well. The specialist will take a detailed history of the person's symptoms and gather information to gain a wider picture of the person's behaviour and functioning in their daily life.

Standard tests of mental abilities, which tend to focus on memory loss, can be less helpful in the diagnosis of FTD. More specialised tests of social awareness or behaviour may be needed.

CT (computerised tomography) and MRI (magnetic resonance imaging) scans of the brain should be used to assess the pattern of damage. They can also rule out other possible causes of a person's symptoms, such as a stroke or tumour. If further tests are needed, more specialised brain scans will be carried out, such as PET (positron emission tomography) and SPECT (single photon emission computerised tomography) to measure the person's brain activity. These scans are useful as they may detect reduced activity in the frontal and/or temporal lobes before a CT or MRI scan can detect structural changes to these lobes. Further tests may include a lumbar puncture, which involves collecting and analysing fluid from the spine and is carried out mainly in younger people.

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Where a person is suspected of having a strongly inherited form of FTD, genetic testing may be able to confirm the diagnosis. This could then allow family members to find out whether they will go on to develop the condition in their lifetime. The decision to find out is up to the individual and support is available.

After a person dies, it is possible to make a pathological diagnosis of FTD, as the changes to the brain can be directly seen at a post-mortem.

Treatment and support

Researchers are working to find effective new treatments for FTD, but there is currently no cure and the progression of the disease cannot be slowed. Treatment tends to focus on helping the person live well by easing their symptoms and supporting them and those around them.

Supporting a person with FTD usually requires input from a team of professionals. These can include a GP, community nurse, psychiatrist and speech and language therapist. When someone has problems with movement or co-ordination, support from a neurologist, physiotherapist or occupational therapist is often needed as well.

Caring for someone with FTD can be particularly challenging, because of the person's age and the changes in behaviour and communication.

Specialist support groups for younger people with dementia or those with FTD, as well as their carers, can provide invaluable practical and emotional support (see 'Other useful organisations' for details, including specific support groups for people affected by genetic FTD). Social interaction can also help if the person seems to lose motivation in things or appears bored or lonely.

If a person is found to have a gene mutation that causes FTD, birth relatives will also have to decide whether to have genetic counselling and testing themselves. Testing unaffected family members can be a cause of great anxiety. The result can have emotional, psychological, social, practical and occupational implications. This is why testing would only be done after extensive counselling with a geneticist.

Behavioural changes

Many people with FTD continue to lead an active social life for some years following diagnosis, but changes in their behaviour can begin to make social situations more challenging. It can be less stressful for carers if they try to accept awkward and potentially embarrassing behavioural symptoms as part of the disease, rather than confront or correct the person, unless the behaviour poses a risk of harm.

The person with dementia will generally lack insight into their condition or the impact of their behaviour on others. They will also generally not have much control over their actions. When a person with FTD behaves inappropriately in public, it can be useful for the carer to try to remove any triggers for this behaviour, or distract the person with something else.

Some carers of people with dementia carry a small card that explains to members of the public that the person has dementia. Alzheimer's Society produces helpcards that you can use for this purpose. To order go to alzheimers.org.uk or call **0300 303 5933**.

Problems with lack of insight and impulsive behaviour make safe driving very difficult for someone with FTD. Driving is often a very sensitive issue that needs careful handling. For more on this see factsheet 439, **Driving and dementia**.

It may be easier for a carer to allow the person to carry on with other behaviours, as long as they are harmless. The person may prefer to follow a fixed routine or pursue an obsession (eg with jigsaws or music), and it may be best to let them. However, some of these behaviours, such as compulsive eating and drinking, won't be harmless. Many carers try to help minimise a person's opportunity for compulsive eating – for example, by offering food only at mealtimes and in suitable portions or healthier (eg low-fat) options. The person's use of alcohol may also need to be closely monitored.

It is important to try to manage restlessness, agitation or aggressive behaviour without drugs initially, where possible. This behaviour might result from a person trying to communicate an unmet need, such as feeling frustrated or in pain. Physical exercise and enjoyable, tailored activities, carried out as part of a routine, can help to reduce these types of behaviour.

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There is evidence that certain antidepressant drugs can help some people with FTD feel less apathetic (having little interest in people and things) and relieve some behavioural symptoms.

If antipsychotic drugs are being considered for a person with FTD, it is recommended that a specialist advises on the risks and benefits.

There have been a few small trials of drugs used to treat Alzheimer's disease (donepezil, rivastigmine, galantamine and memantine) in people with FTD. These have had mixed results. In some cases, these drugs made people's symptoms worse. They are also not licensed for use in FTD and are not widely prescribed.

Language problems

A speech and language therapist with the right experience will be able to support someone with FTD who is gradually losing their language skills. They will try to maximise a person's existing skills and find new ways for them to communicate.

A therapist can advise a person's carer on new ways of listening and talking – for example, talking in simpler sentences. In time, a person who is losing their language skills may be taught non-verbal ways of communicating. These can include the use of gestures, drawing or electronic devices.

A speech and language therapist can also help if a person has problems swallowing and can offer practical advice on eating and drinking.

Other useful organisations

Motor Neurone Disease Association

PO Box 246
Northampton NN1 2PR

01604 250505
enquiries@mndassociation.org
www.mndassociation.org

National charity that funds and promotes research into motor neurone disease and provides support for people affected.

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PSP Association

PSP House
167 Watling Street West
Towcester
Northamptonshire NN12 6BX

300 011 0122
helpline@pspassociation.org.uk
www.pspassociation.org.uk

Offers advice, support and information to people living with progressive supranuclear palsy and corticobasal degeneration. Also supports research into treatments and ultimately a cure for these conditions.

Rare Dementia Support

Box 16, National Hospital for Neurology and Neurosurgery
Queen Square
London, WC1N 3BG

020 3448 4773
www.raredementiasupport.org

Runs specialist support group services for individuals living with, or affected by, one of five rare forms of dementia: frontotemporal dementia, posterior cortical atrophy, primary progressive aphasia, familial Alzheimer's disease and familial frontotemporal dementia.

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Reviewed by: Prof Jason Warren, Senior Clinical Fellow and Professor of Neurology and Jill Walton, Nurse Advisor, Rare Dementia Support, Dementia Research Centre, Institute of Neurology, University College London

This factsheet has also been reviewed by people affected by dementia.

A list of sources is available on request.

Alzheimer's Society National Dementia Helpline England, Wales and Northern Ireland:

0300 222 1122

9am–8pm Monday–Wednesday

9am–5pm Thursday–Friday

10am–4pm Saturday–Sunday

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