# General Information on FTD

(Frontotemporal Dementia)

**ALZHEIMER'S AND RELATED DISORDERS SOCIETY OF INDIA**
Hyderabad Deccan

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Material taken from: **Frontier - Frontotemporal Dementia Group**

# General Information on FTD

Frontotemporal dementia, sometimes called frontotemporal lobar degeneration, was first described 100 years ago by Arnold Pick and was previously referred to as Pick's disease. It is the second most common degenerative disease causing dementia in younger adults. The age of onset is typically in the 50s or 60s but can be as young as 30.

Damage to brain cells is more localised than in Alzheimer's disease, and begins in the frontal and/or temporal lobe. In FTD the clinical presentation varies, depending on whether the frontal or temporal lobe is affected first. When the initial pathology affects the frontal lobes, the main changes are in personality and behaviour (**Behavioural-variant FTD**). Individuals with predominant temporal lobe involvement present with loss of language skills (**Progressive Non-fluent Aphasia**, **Semantic Dementia**) known as progressive aphasia (aphasia is the loss of the ability to produce or understand language).

The pathology of FTD is much more complex and variable than the pathology of Alzheimer's disease. Instead of the 'plaques and tangles' which characterise Alzheimer's disease, the brains of people with FTD brains show a severe loss of brain cells (neurons). In some individuals, the tau protein, which is also involved in Alzheimer's disease, collects in neurons known as 'Pick bodies'. Asmall proportion of people with tau accumulations have a mutation of the tau gene on chromosome 17. More commonly, the brains of people with FTD shows an accumulation of another cell protein ubiquitin. Ubiquitin is involved in clearing waste products from brain cells but for reasons that are currently unknown, this protein builds up in some people with FTD. Very recent research has suggested that the accumulation of ubiquitin is attached to another protein (called TDP-43) which has a fundamental role in cell nuclei. Again, a small proportion of people with ubiquitin accumulation have a genetic mutation, this time of the progranulin gene which is also located on chromosome 17.

# Deficits in Frontotemporal Dementia

Deficits associated with frontotemporal dementia are varied and depend on the location and severity of pathology in the brain. Most common deficits are changes in behaviour and personality, difficulty relating to other people and difficulty organising day□to□day activities. In these patients, the underlying brain changes affect predominantly the frontal lobes (**Behavioural- variant FTD**). In contrast, other patients show change in language proficiency, either in the form of a difficulty understanding the meaning of words (**Semantic Dementia**), or a difficulty using the correct words (**Progressive Non-fluent Aphasia**) . This may be accompanied by a difficulty judging emotional state in self and others accurately. In these patients, the pathology is more pronounced in the anterior portion of the temporal lobe, or in the region where the frontal lobe meets the parietal and temporal lobes. Over time, and as the disease progresses, pathology tends to become more diffuse. As a consequence, clinical presentations tend to merge and deficits become more pronounced.

# Managing Frontotemporal Dementia

Not uncommonly, individuals with frontotemporal dementia show a limited awareness of their deficits in thinking and changes in behaviour and a reduced understanding of the impact of their condition on friends and families. This limited insight may create significant challenges for

carers. Although each situation is unique, a number of simple guidelines may be helpful. First, carers need to recognise that behavioural changes are the result of changes in the brain. In most instances, the person is not being difficult intentionally. As such, direct confrontation to change a difficult or inappropriate behaviour is unlikely to be successful. You are more likely to succeed by changing the environment, for example by redirecting attention or removing what triggered the behaviour, than by trying to change the person.

In individuals where language deficits are the most prominent symptoms, difficulty understanding others or being understood by others may result in frustration. Simple, rather than complex, commands or instructions and the use of simple words are likely to improve comprehension. Use of visual supports, such as drawings or photos, may also be helpful in situations where verbal expression is disrupted.

# Behavioural-variant FTD

In the frontal or behavioural variant of frontotemporal dementia, the person's mood and behaviour may become fixed and difficult to change, making individuals appear selfish and unfeeling. A loss of empathy and emotional warmth is very common. In contrast to Alzheimer's disease, recent memory is typically preserved.

Apathy or lack of motivation is very common, leading people with FTD to abandon hobbies and avoid social contact. Others lose normal inhibitions and start talking to strangers or exhibiting embarrassing behaviour in public.

Difficulty in reasoning, judgement, organisation and planning is frequent, along with a reduction in spontaneous conversation. Changes in eating patterns are very common often with a craving for sweet food, a tendency to overeat and a restriction in food preferences.

A decline in self-care and a reduction in the ability to perform activities of daily living is another early feature. As the disease progresses, the person may become 'obsessional', repeating patterns of movement and behaviours like handwringing or echoing back whatever is said.

# Semantic Dementia

In patients with the temporal lobe form of Frontotemporal Dementia, the initial symptom is usually a decline in language abilities. The left temporal lobe is critical for the fluent production of words and especially for assigning meaning to words. Because the language disorder associated with temporal lobe pathology reflects a breakdown in the meaning (or semantic) system underlying language, we prefer the term semantic dementia which conveys the core deficit and progressive nature of the disorder.

Patients typically complain of a “loss of memory for words” involving, at first, less common words and particularly the names of peoples'. As the disease progresses impairment of word comprehension is noticeable which again affects less common words. Reading and spelling are also typically affected, although numerical abilities can be remarkably well spared.

If the right temporal lobe is involved then patients (or carers) often notice problems recognising previously familiar people. It is not uncommon for patients to talk to people as if they were strangers only to discover later that were old friends.

Day-to-day memory is relatively spared but may appear poor due to difficulty with expression. In later stages, the disease spreads to the frontal lobes so that many of the features listed above, especially the changes in emotional responses, empathy and food preference. We have noticed that many patients with semantic dementia are extremely good at puzzles, bingo and jigsaws, and retain sporting abilities such as bowls or golf until very late in their illness.

Skills associated with posterior brain regions such as navigation, route finding and eye-hand coordination are spared at least until very late in the disease.

## Progressive Non-fluent Aphasia

Progressive non-fluent aphasia (PNFA) is the least common form of Frontotemporal Dementia and affects the ability to speak fluently. Patients present with difficulty communicating due to slow and laboured production of words often with distortion of speech and a tendency to produce the wrong word.

Some patients have slurring of speech whereas others are able to articulate words but produce frequent near misses (e.g. they say “silter” for “sister”). Understanding of word meaning is preserved, but patients with PNFA have problems comprehending sentences and following conversations, especially if there are a number of speakers. Using the telephone and communicating with groups of people is particularly difficult.

Spelling is frequently impaired from an early stage and some patients develop difficulties reading (so called dyslexia). Subtle deficits in problem solving, mental flexibility and decision making are often present from an early stage. Changes in behaviour are uncommon in the early stages but do occur later.

Some people develop clumsiness of handuse, known as apraxia. In later stages, the disease spreads to the frontal lobes, so that many of the features described above, especially the changes in emotional responses and empathy occur. There is considerable overlap between progressive nonfluent aphasia and corticobasal degeneration (see section on *Overlap with Corticobasal Degeneration*).

## Is treatment possible?

There is no cure and no effective disease-modifying treatment yet available, but various therapies can help some of the symptoms such as aggression or overeating.

- Psychological methods are very important to help control and contain abnormal behaviour.
- Caregiver support and counselling is vital.
- All families should receive expert advice on genetic aspects.
- Speech therapy is of benefit in patients with progressive non-fluent aphasia, particularly helping to develop alternative communication methods.
- The input of an occupational therapist can be invaluable to improve everyday functioning at home.

# Frontotemporal Dementia in India

FTD is a relatively under-recognised problem in India. During the past few years however, there has been a gradual increase in patients and families, who are identifying the problem and seeking medical help. Doctors in India have begun to diagnose FTD and it has been found that about 15% of dementia patients who are seen in Memory Clinics have FTD. All three subtypes of FTD are seen- Behavioural variant FTD is the most common, followed by Progressive Nonfluent aphasia and Semantic Dementia. Some patients develop signs of Parkinsonism during the course of disease and about a quarter of them have a family history of FTD. Pathological confirmation of tau protein deposition in the brain has been demonstrated in few patients. Genetic testing is being developed in a few research centres and is not widely available

Clinical experience with Indian patients suggests similar behavioural problems to those seen in their western counterparts, but culturally specific differences appear to exist. The experience with FTD patients is that they seek medical attention quite late in the disease, after causing considerable stress to families. In part, this is because abnormal social behaviour is not recognised to signify brain disease and awareness is only now increasing.

# Challenging behaviours: managing tips

People with dementia may develop behavioural symptoms that may be a consequence of the person no longer being able to meet his or her own needs, or be a direct symptom of changed brain function. Behavioural symptoms vary between individuals and change over time as the illness progresses. If behaviour is having an impact on the person with dementia or those around them, consider the following:

**Know what functions of the brain have been affected**
To manage behavioural symptoms the family carer needs information. This includes finding out from the medical specialist or GP what functions of the brain have been affected by the illness. If the person with dementia has seen a neuropsychologist he or she will also be able to give this information.

**Know the person**
It is important to know the person well. This includes his or her personality, past experiences, likes and dislikes, and the things which are important to him or her. The family carer is best placed to know this information and use this knowledge when developing behavioural management plans.

**Ensure the person with dementia is as physically well as possible**
When people with dementia are unwell they will be less able to use the skills they still have. Having check ups with the GP, providing good nutrition, encouraging exercise and managing medications will assist with this. Check for visual and hearing problems and make sure the person has glasses or hearing aids if needed. If behaviour deteriorates rapidly see your GP as there may be a medical cause for this (such as urinary tract infection).

**Manage/examine your own behaviour**
Behavioural symptoms in people with dementia are made worse when their family carers are stressed. A high proportion of carers develop significant depression which further lowers the ability to deal with the person with dementia.

**Modify the environment**
The environment has an impact on the ability of the person with dementia to use his or her skills. An environment which has cues for people with memory loss and disorientation will assist in meeting their needs more effectively and can reduce frustration or fear. A noisy and over-stimulating environment may interfere with concentration and comprehension leading to more agitation.

**Reflect on incidents**
Reflecting on what is happening can be a useful learning tool. Think about possible triggers that have promoted the well-being of the person and triggers that contribute to the person's behavioural symptoms.

# Apathy or lack of motivation

Remember this is not laziness. The brain is not initiating the activity for the person. You need to be the initiator.

**Managing Apathy**

- Don't try to reason with person (this is limited due to inability to understand consequences)
- Prompt the person to do the activity
- Encourage the person
- Start activity for them
- Mirror activity (physically show the person what you want them to do)
- Don't take over activity

# Compulsive Behaviour

**A person with FTD may:**

- Check locks and doors over and over
- Have rigid walking patterns
- Hoard items
- Count over and over
- Go to toilet frequently

**Managing Compulsive Behaviour**

Is it a problem or can it be ignored?

- Distract

- Capitalise by using compulsion as activity when possible

# Aggression

May be more common due to the brain's inability to control impulsive actions.

**Preventing Aggression**

- If there is an incident of aggression think about what may have triggered it and how you might avoid the situation again.
- Ensure person does not have pain & see GP for illness.
- When talking to the person do not use sarcasm or abstract thinking i.e. be concrete. Reduce external distractions when talking e.g. TV/radio or in busy places.
- Maintain routine when possible. If necessary make changes gradually
- Don't rush the person

**Managing Aggression**

- Maintain your and others safety
- Do not argue or rationalise
- Leave if possible and try again later
- Empathise (may or may not respond)
- Distract
- Keep calm, lower your tone of voice, be aware of body language (may or may not be recognised by person with FTD but will assist in calming you)

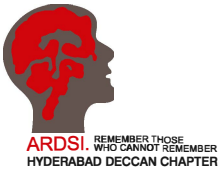
# Eating Problems

People with FTD may change their eating habits. They may change the types of food they want to eat e.g. sweet foods, eat the same foods or have odd combinations of food. They may 'cram' their food or eat food from other plates.

**Management**

- Monitor the amount of sweet food to prevent excessive weight gain. Limit amount available in house.
- May substitute with 'diet' food. Encourage sweet food which has nutritional value e.g. fruit.
- If 'cramming', provide small portions at a time, or remove dishes from the table when possible.

If possible, tell your friends in advance when eating together, so that you do not feel embarrassed. You will be surprised how understanding some friends can be.

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