

CFU Carers' Support Group

For carers and relatives of people with fronto-temporal dementia and semantic dementia

Newsletter

FEBRUARY 2005

Welcome

Welcome to the current edition of our newsletter, which follows the third meeting of the carers' support group earlier this month.

We'd like to offer a big thank you to all of you who managed to make it through the traffic on that day. We were glad to see that so many of you were able to come at the alternative time of 3pm.

Once again, the meeting was a big success, with a warm and friendly atmosphere. Professor Neary gave an excellent talk, explaining fronto-temporal dementia and semantic dementia in detail.

We were pleased that so many of you had questions to ask at the end of the talk, and hope that you found the meeting useful.

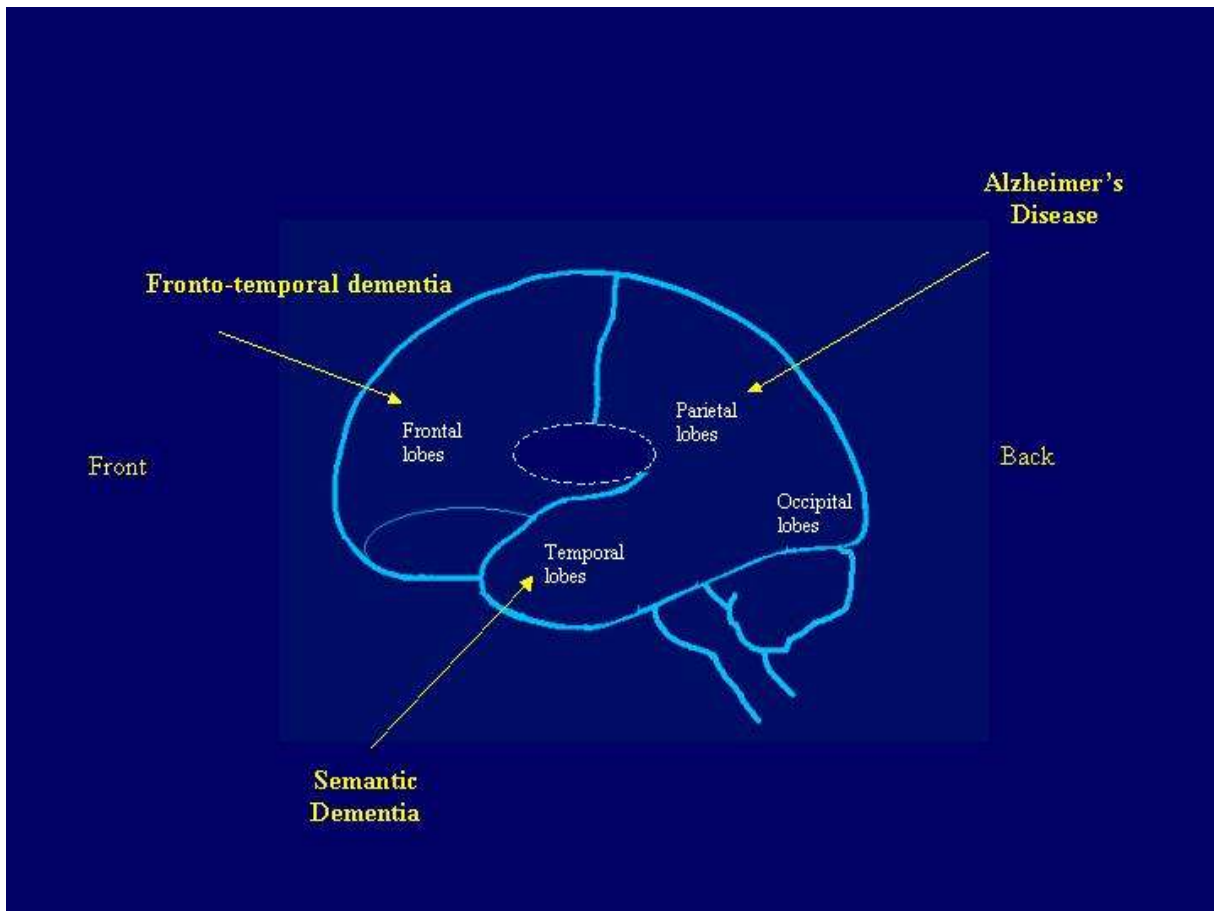
For those of you who were unable to attend, we've summarised the main points of Professor Neary's talk, including some of the questions that were asked by carers. We hope that you will find the information interesting and helpful. Feel free to write in with any other questions, and we will try to answer them in our next newsletter.

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Functions of the Brain

To get a good understanding of fronto-temporal dementia and semantic dementia, it is useful to look at the brain and the way it functions. Here's a picture of the brain for you to refer to:



The different areas of the brain play different roles. The posterior areas (i.e. the parietal and occipital lobes) are responsible for our visual functions. They help us to see **what** things are and **where** they are in space. The temporal lobes play an important role in memory and language. They enable us to **learn** and **store** new information. Finally, the frontal lobes are responsible for **organisation** and **control** of information. They enable us to generate plans and make decisions. This is often referred to as '**executive function**'.

Dementia

Traditionally, 'dementia' has been viewed as a singular, global decline of mental functions. However it is increasingly clear that this is not the case. Different disease processes affect specific parts of the brain, resulting in diverse patterns of mental decline. The location of the disease determines the symptoms that occur. As you can see on the diagram (p. 3), fronto-temporal dementia and semantic dementia affect areas towards the front of the brain, whereas Alzheimer's disease affects the areas at the back. The different disease processes are therefore associated with different symptoms.

Patients with Alzheimer's disease typically have problems in memory, visual and spatial function, and language. They find it difficult to learn new information, see things around them, and retrieve words in conversation. In contrast to their often debilitating mental decline, they are fully insightful and concerned about their problems, with preserved personality and behaviour.

As you will be aware, patients with fronto-temporal dementia and semantic dementia

are entirely different. Since the posterior areas of the brain remain unaffected, there are no problems in visual function. However, the affected frontal areas are responsible for controlling behaviour, meaning that there are often profound changes in personality.

The underlying disease

Fronto-temporal dementia and semantic dementia form part of the same disease, known as '**fronto-temporal lobar degeneration**'. The underlying disease process is the same. It is the **location** of the disease that determines the symptoms and the corresponding name that is given to the dementia. As you can see in the earlier picture, in fronto-temporal dementia, the disease primarily affects the frontal lobes, whereas in semantic dementia the disease selectively affects the temporal lobes. The two disorders will be described separately to illustrate the differences between the symptoms.

Fronto-temporal dementia

Fronto-temporal dementia (FTD) typically affects people below the age of 65. About half of the patients that we see report a family history of similar problems, and in a

small proportion of these cases, there is a specific genetic fault associated with developing the disease. However, FTD also occurs in people who have no other affected family members.

Symptoms

Patients are typically physically fit. Initial changes are apparent in mental function only. As the disease progresses, reduced activity and physical slowing may occur.

Symptoms arise gradually, making it difficult to pinpoint the very beginning of the disease. Carers often report with hindsight that their relative showed signs of depression, anxiety, over-sentimentality, or hyperchondriasis even before they noticed clear changes in behaviour. Commonly, referral to doctors occurs following difficulties at work, or social problems within the family.

As you will be aware, FTD principally affects **personality** and **behaviour**. You may have observed some of the following:

- Irritability
- Egocentricity/selfishness
- Inappropriate/disinhibited behaviour
- Lack of embarrassment

- Loss of feeling for others
- Poor insight into problems
- Loss of interest in former hobbies
- Apathy and need for prompting
- Restlessness/wandering behaviour
- Mental rigidity/inflexibility
- Obsessive/ritualistic traits
- Odd mannerisms
- Preoccupations
- Poor personal hygiene
- Changes in eating behaviour

Symptoms are not exactly the same for all sufferers, and depend on which part of the brain is most affected. Generally speaking, there appear to be two different patterns:

Apathetic: Patients are inert and inactive, and act as if they are depressed. However, they do not complain of feelings of depression, and remain unconcerned about their lack of motivation and drive.

Disinhibited: Patients are overactive and act inappropriately in social situations. They show a lack of concern for the feelings of others and are often restless and irritable.

In addition to changes in personality, you may have noticed some problems in

memory, language, and ability to perform daily activities.

Memory

Sufferers often have problems in concentration, which in turn can affect memory ability. You will know from your own experience that if you are not paying attention to or concentrating on information that is being given to you, you will not remember it afterwards. This appears to be the reason why FTD patients remember some things and not others – it is not that they ‘forget’ information, rather that they fail to take it in when it is presented.

Language

Spoken output may reduce gradually, and eventually, sufferers may become totally silent. Patients may also develop ‘verbal stereotypies’, repeating the same phrase or topic over and over again. Sometimes patients may use the wrong words when talking – this is related to the temporal lobes, and will be discussed further in the ‘semantic dementia’ section.

Performing everyday activities

Patients may have problems in sequencing and ordering task components, generating

appropriate strategies, and grasping instructions. In everyday life, this may be reflected by reduced ability to perform seemingly simple tasks, such as putting clothes on in the right order, or using household gadgets. This is related to the failure of the frontal lobes in controlling and organising information.

Semantic Dementia

Semantic dementia (SD) is another clinical manifestation of frontotemporal lobar degeneration, associated with shrinkage of the temporal lobes (see diagram on p. 3). SD is a relatively rare disorder and is much less common than FTD. However, like FTD, people with SD tend to be affected at a relatively young age, generally between the ages of 50 and 65. Relatives report a history of a similar disorder in the family in about a quarter of cases.

Symptoms

In SD, people progressively lose their ability to remember the meaning of words, objects and faces. Changes may also occur in people’s behaviour, although these are qualitatively different from those observed in FTD.

Physical signs

Sufferers are generally fit and physically healthy although they may become slowed down in the later stages of the disease.

Language

Problems in language tend to occur first. People with SD have difficulty naming and often use the incorrect word in conversation (e.g. calling a lion a "dog" or saying "water" for milk). They have difficulty understanding the meaning of words and may fail to recognise relatively simple words in conversation. Sufferers may also repeat the same expression (verbal stereotypy) over and over again (e.g. "oops a daisy", "Well, it's one of those things"). Nevertheless, their speech is generally fluent and effortless, which may initially give relatives the impression that they do not have any primary difficulty with language. Moreover, their repetition is good, and they can generally read and write words they do not understand.

Visual recognition

The disorder may also affect the ability to recognise objects and faces. The sufferer sees normally (and so can distinguish when two objects or faces are the same or different), but is no longer able to attribute

any meaning to what they see. This is why sufferers may still be able to draw accurately objects that they no longer recognise. Occasionally, difficulties in visual recognition may occur before language problems.

Memory

Sufferers do have problems with memory, but it is with a specific type of memory called "semantic memory". There are two different types of memory which are processed in different areas of the brain: "personal memory", which corresponds to memory for day-to-day events, such as remembering what you ate for dinner yesterday or where you went on your last holiday; and "semantic memory", which relates to your general knowledge about the world and, more specifically, to what words and objects mean and represent. In SD, people's personal memory is well preserved, so they have no difficulty keeping appointments or remembering what they have done in the past few weeks. However, they do have a problem with their semantic memory, which means they cannot remember the significance of words, objects and faces.

Semantic memory does not break down in an all-or-none fashion and people with SD typically retain partial knowledge of objects and words. Interestingly, sufferers tend to retain information about things that are personally relevant to them in their everyday lives. For example, they may be able to recognise their own kettle but are unable to make a cup of tea with someone else's kettle because they do not understand its function anymore. Similarly, they will be better able to recognise names and faces of people they see regularly (e.g. spouse, visiting health worker) than of people they only see occasionally (e.g. relatives at a family function). This "autobiographical" effect even extends to words and place names: people are more likely to understand names of countries and cities they have visited recently (e.g. on holiday) than places they have little personal experience of.

Individual differences

Although the disorder affects several domains, such as words, objects and faces, people do vary in the extent to which these domains are affected. Some sufferers have particular difficulties in word meaning and only develop problems in recognising faces later on in the course of the illness. For

other people, the main problem lies in the visual domain, and they may be unable to recognise people's faces at a time when they can still understand their names.

These individual differences between word meaning and visual recognition reflect the location of shrinkage in the brain. Although both temporal lobes of the brain are affected in SD, one side may become more heavily affected than the other. Shrinkage of the left temporal lobe is associated with better recognition of faces / objects and relatively poor understanding of word meaning, whereas predominant shrinkage of the right temporal lobe is associated with poor face recognition and better understanding of words.

Behaviour

Difficulty in comprehension is by far the most important feature of SD, however, sufferers do also show changes in their behaviour. These changes are generally more subtle than those observed in FTD, and have a more obsessional quality. Sufferers tend to focus on a restricted range of activities (e.g. house work, sewing, walking), which they pursue obsessively. They develop a preference for fixed routines and often become preoccupied by time, so that certain

activities are always carried out on set days and at fixed times. Certain people also become excessively parsimonious and may hoard their money in different places

around the house. Sufferers generally become more self-centred although they rarely show the marked loss of empathy and lack of self-care typical of FTD.

Carers' Questions

1. Can FTD or SD be brought on by a head injury?

No. FTD and SD cannot be caused by any environmental factors, such as a head injury, a virus or exposure to chemicals. However, because head injuries generally 'shake up' the brain, they may accelerate the development of emerging symptoms, and difficulties that were already there become more noticeable. This often gives relatives the impression that the head injury was the cause of the disease.

2. Do FTD and SD turn into Alzheimer's disease as the disease progresses?

No. Alzheimer's disease and FTD / SD are two separate diseases, which affect different parts of the brain and so lead to different patterns of symptoms. As described above, Alzheimer's disease affects the back part of the brain and is associated with difficulties in memory, language, and visual and spatial function. As the disease progresses, the symptoms become worse but do not change in nature because the front parts of the brain remain spared. The same is true for FTD and SD. Because the back areas of the brain are not affected in these diseases, people do not develop the same problems Alzheimer's sufferers and have no difficulty with their vision or finding their way around their environment.

3. Why don't treatments for Alzheimer's disease work in SD and FTD?

The brain produces a range of chemical substances (called "neurotransmitters") to transport messages from one part of it to another. If the amount of these chemicals is reduced, then the areas of the brain that are dependent on them slow down and do not work as efficiently. Although this does not explain the whole problem, we know that people with Alzheimer's disease have reduced levels of a neurotransmitter called 'acetylcholine', and that this contributes to their symptoms. Treatment for Alzheimer's disease slows down the progression of symptoms by boosting the levels of acetylcholine in the brain. However, people with FTD and SD have normal levels of this substance in their brain and so do not benefit from treatment for Alzheimer's disease. We now know that FTD and SD are associated with deficiency of a different neurotransmitter, called serotonin. There is some evidence that boosting levels of serotonin in some people can help with certain disruptive, repetitive behaviours, however the disease process appears to be more complicated than this and such treatments are by no means miracle cures.

4. Can sufferers develop both FTD and SD?

Although the two diseases are related, most people's symptoms stay the same and remain well circumscribed. Occasionally, patients may develop certain features characteristic of the other disease. For example, FTD sufferers may develop mild difficulties in understanding word meaning or SD patients may become a little apathetic in their behaviour as the disease progresses. However, these changes are relatively mild and there is never a complete crossover between diseases. That is, FTD does not change into SD and vice versa.

5. Are FTD and SD inherited diseases?

In the majority of cases, the disease occurs sporadically, that is, without any history of a previous similar illness in the family. However, occasionally, it may occur in more than one member of a family, suggesting a genetic predisposition in some people. Families who have a genetic predisposition towards FTD or SD are generally already aware that the illness runs in the family, because they know of other relatives who have developed similar symptoms. It should be emphasized that these cases are very rare and should not be regarded as the norm.

6. Are there any physical symptoms in FTD and SD?

People with FTD and SD are generally fit and healthy and do not have any physical symptoms. Many of our patients continue to exercise and pursue active hobbies for many years. In the late stages, people may slow down and show symptoms akin to a mild form of Parkinson's disease ("parkinsonism"). They slow down physically and may develop problems in balance and slight trembling of the limbs.

8. Is sense of smell affected in SD?

Yes. It's not only the meaning of words and objects that is affected in SD. Sufferers also have difficulty recognising everyday sounds, tastes and smells. Although their sensory abilities are well preserved (i.e. they can distinguish between two different sounds or smells), they have difficulty ascribing a meaningful identity to what they perceive and so may be unable to recognise familiar sounds, such as the ringing of the telephone or doorbell, or identify the smell of cooking as a sign that food is being prepared.

9. Are sufferers aware of pain and temperature?

An altered reaction to pain and touch is frequently observed in FTD and SD. Certain sufferers react in an exaggerated manner to non-threatening stimuli, such as a light touch on the shoulder, and yet may show a reduced response to pain and not even flinch when they have visibly injured themselves. Similarly, they often seem to be less aware of extremes of temperature and may step into a scalding bath without realising they have burned themselves.

7. Can language problems be helped?

In FTD, lack of communication is due to a person's gradual loss of social awareness rather than to any true difficulty with language. Because of progressive damage to the frontal lobes, patients have difficulty seeing a situation from someone else's point of view and so do not respond to other people's conversation. In these cases, stimulating someone by talking to them a lot may not be of much help, because the sufferer will not understand the point of it. In SD, The sufferer's difficulty is in understanding the meaning of individual words. However, it can often help understanding if words are put into a context: speaking in complete sentences is better than communicating in short phrases or by using single words. As we saw above, patients generally retain understanding of things that are personally relevant to them. Using words and expressions that the sufferer himself/herself uses in everyday life will help their understanding in conversation.

10. Can the sufferer alter his/her behaviour?

Although it may sometimes feel like sufferers are acting in a deliberately awkward or selfish way, their behaviour is the result of their brain disease, and is not under their control.

Patients are no longer able to see things from someone else's point of view, so are unaware of the effect of their behaviour on others.

11. Do all FTD sufferers ultimately become disinhibited?

No. Although there are two main patterns of behaviour in FTD, apathetic behaviour and disinhibited behaviour, all patients eventually become more apathetic as the disease progresses.

People with apathetic behaviour do not become disinhibited.