

CFU Carers' Support Group



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

Newsletter

AUGUST 2008

Welcome

Welcome to the August edition of our CFU Support Group Newsletter. Thanks to all of you who came to our carers' meeting on 31st July. Dr Anna Richardson, one of the Consultant Neurologists from the Cerebral Function Unit clinic gave a really interesting and helpful presentation about the physical aspects of frontotemporal dementia and semantic dementia. This was part of our rolling programme of talks, aiming to give

information about every aspect of the disorders. We've provided a short summary of the main points of the talk and also some of the discussion points that group members contributed. We hope you find the information useful and if you have any additional comments or questions, try the website, where you can find previous editions of the newsletter, summaries of other talks, and other useful information and links:

www.cerebralfunctionunit.co.uk/carers.html

Housekeeping notes:

Contributions:

We are always happy to hear your comments on the carers group and would appreciate your guidance on how to develop it. What are *your* needs? What do *you* need to know about? What kind of people would *you* like to hear from? If you have any suggestions for speakers, please let us know. Also, if you would like to share your experiences with the group, at a meeting, or by writing to the newsletter, we would be very happy to help.

Donations:

The CFU Carers Support Group is run on a purely voluntary basis and receives no external funding. We greatly appreciate the donation of £1 from carers who attend the meetings. This goes towards providing teas and coffees at the meetings and offering a small gift to invited speakers who kindly give their time to come and speak to our group.

Physical aspects of frontotemporal and semantic dementia

Dr Anna Richardson, Consultant Neurologist, Cerebral Function Unit, Greater Manchester Neuroscience Centre

Introduction

Talking about physical problems in frontotemporal dementia (FTD) and semantic dementia (SD) may initially seem rather odd as the conditions are known primarily to affect mental rather than physical health. However, physical problems do tend to occur in the later stages, and also in rarer forms of the disease.

A recap of brain functions and scientific terms

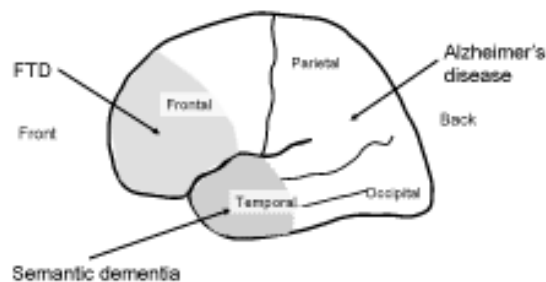
Before getting into the details, it is important to recap on the functions of the brain and the scientific terms involved.

The term 'frontotemporal lobar degeneration' ('FTLD' for short) is an umbrella term for the unifying disease process which causes a number of disorders, including frontotemporal dementia (FTD), and semantic dementia (SD).

To get a good understanding of these changes in frontotemporal dementia and semantic dementia, it is important to look at the brain and the way it functions (see diagram). The areas at the back of the brain (the 'parietal' and 'occipital' lobes) process the information that comes in through our senses, and are important for vision and spatial navigation. In contrast, the 'frontal' and 'temporal' lobes act as a control centre, and help us to integrate and make sense of this information. This is very important in our understanding of the meaning of things. It is these parts of the brain that are affected in FTD and SD. They cover the largest area of the brain in humans, and develop later in childhood. It is therefore unsurprising that you might have noticed that the person you care for acts in a 'childlike' fashion – the parts of the brain that have not yet developed in children are the same as those that are not working in people with these conditions.

Frontotemporal lobar degeneration is a brain disease which generally affects the front parts of the brain. In most people, it affects either the frontal or the temporal lobes. If it affects mostly the frontal lobes, the syndrome of frontotemporal dementia occurs, associated with personality and behavioural change. If the temporal lobes are mostly affected, this gives rise to the syndrome of semantic dementia, associated with a loss of understanding of the meaning of words and objects. In some cases, FTLD may affect both frontal and temporal lobes similarly, and the person will exhibit a mixture of both FTD and SD syndromes.

Diagram of the brain: the areas affected by FTD and SD



In the following sections, we will use the term Frontotemporal Lobar Degeneration (FTLD) to talk about the disease in general.

Physical problems secondary to behavioural disorder

People with FTLN generally undergo marked changes in personality and behaviour, into which they have little or no insight. These changes can impact upon their physical activity in different ways.

Their behaviour tends to take one of two forms: overactive, or apathetic. People with the 'overactive' type of behaviour are often disinhibited and restless and often pace around repetitively without cause. Those with the 'apathetic' type of behaviour typically show a decline in physical activity and will often spend all day in bed or sitting still in an armchair, showing no desire to go out. It is often difficult to persuade them to go out of the house at all. In both types, people actually remain physically well. However, the changes may nonetheless be interpreted as a physical problem. For example, a person's loss of mobility due to their apathy and poor motivation may be viewed as a physical difficulty, even though, in reality, the person is fully able to walk.

Similarly, repetitive behaviours may develop, such as rubbing of the hands, arms or legs. This is not because these areas are physically hurting – it is simply a repetitive action that is part of the behavioural change. People with FTLN also sometimes 'pick' at their skin – this again tends to result from repetitive behavioural actions, rather than a physical skin complaint.

Incontinence

Incontinence does affect some people with FTLN but by no means affects everybody. Again this is not due to a physical or neurological problem but can occur secondary to the behavioural change. People who are apathetic can exhibit a lack of self care and may neglect their personal hygiene and appearance. Incontinence may arise because they are no longer embarrassed by their personal appearance in public. People with FTLN may no longer understand the social norms and conventions and therefore fail to follow them. Although incontinence in this case is not a physical problem, it can be helped by using the same medication that is used to treat people with physical bladder problems.

Later stages of the disease – parkinsonism

People in the late stages of FTLN generally experience some physical problems, such as stiffness and slowness. These are termed 'parkinsonism' by doctors because they are similar to those seen in Parkinson's disease. Parkinsonism relates to problems in the control of movement, with movements becoming slower, limbs and neck stiffer, and sometimes the development of a tremor (shaking) of the limbs (although this is less common in FTLN). Relatives tend to report that the person is walking more slowly, and with a stooped posture. Sometimes their face becomes less expressive than before.

In FTLN, these symptoms tend to emerge in the later stages of the disease because the disease process is spreading deeper into the frontal lobes to parts of the brain that are responsible for the control of movement. Unfortunately, the medication that is used in Parkinson's disease does not improve symptoms in FTLN as the different disorders have different mechanisms.

Physical problems never seen in FTLN

- Epilepsy/fits
- Muscle jerks
- Sudden changes in physical state: physical symptoms related to FTLN always appear and develop gradually

Rarer forms of FTLD

FTDP-17

Most cases of FTLD are sporadic (not inherited). Only a small number of cases are passed down through families. In these families (who carry the FTDP-17 gene) it is often common to see movement disorders associated with FTD, either in the same individual, or in different members of the same family. Although these cases are rare, these families have been studied extensively by scientists who believe that they may be the key to understanding the underlying genetic basis of all forms of the disease, including sporadic ones.

FTD-MND

Motor neurone disease (MND) is a rare but well known disease of the nervous system. Some patients with FTLD may develop features of MND and present with prominent physical features in addition to the behavioural and cognitive changes of FTLD. Alternatively, some people develop the physical changes of MND first, and then later acquire the changes associated with FTLD. Cases of FTD-MND are very rare.

Questions

My husband has a problem with his swallowing – on one occasion he had some food in his mouth for about four hours and didn't swallow it. Will this get any better?

Swallowing problems in FTLD are not common and are not very well understood. One thing we do know is that there are typically no **physical** reasons for swallowing abnormalities – when tests are carried out on the throat and tongue, there is no evidence to suggest that the 'tools' involved in the swallowing actions are not working. It is more likely that the problems arise due to cognitive difficulties, such as problems in sequencing (i.e. the person cannot sequence the action of chewing the food then swallowing it in small chunks).

Should antipsychotic medication be used for agitated behaviour?

Antipsychotic medications are good at reducing agitation. However they are known to worsen physical symptoms, making it difficult to choose between the benefits of the drug and the costs of the side-effects. It is notable that the newer antipsychotic drugs are better designed than the old ones and have fewer physical side effects.

What is the expected course of the disease after the physical symptoms?

Except for in the rarer forms of the disease, physical symptoms are typically a late feature. As people with FTLD become less mobile, infections are more likely to occur, leaving people more vulnerable to physical deterioration.

My husband tends to show 'selective motivation'. Is this common?

Yes – it is very common for relatives to report this. People with FTD and SD will often only 'do what they want' and refuse to do anything else. Sometimes they can be on their 'best behaviour' when in front of others, making it difficult for you to demonstrate how difficult life can be for you on a daily basis. One of our carers suggested that a possible advantage to this is that you could use the activity that they *do* like to do as a reward for doing something one of the things that they *don't* like to do. Of course, this might not work for everybody, but it could be worth a try.

The person I care for is diabetic and always tries to eat sweet things regardless. The diabetic nurses and doctors don't understand the dementia. Shall I try and get a referral to a homeopathic treatment centre?

Of course, it can't do any harm to go down that route, but maybe it might be useful to give the diabetic clinicians a second chance first. One thing you could do is to send them information from the Cerebral Function Unit clinic, informing them about the condition. The CFU has specially designed leaflets for clinicians that are available on the website:

<http://www.cerebralfunctionunit.co.uk/informationclinicians.html>

Also, you could ask the consultant neurologist that you see to send a letter, giving them more details about the condition and explaining why they are not being compliant with their diabetic diet and treatment. Hopefully this will help the diabeticians to understand the dementia more clearly.

Will the new drug for Alzheimer's disease (Rember) help people with FTD?

Unfortunately, as it is a different disease, with different pathology, Rember will not help in the majority of cases of FTD. However in the rarer 'FTDP-17' type, people develop 'tangles' in their brain, similar to those in Alzheimer's disease, so possibly it could help in those cases. Nonetheless, this is a very new drug, and is still in the early trial stages.

My mum is always complaining of 'feeling ill'. Is this part of the dementia? She's been tested for lots of other illnesses and all the tests have been negative.

Many people with FTD complain of feeling 'ill' or headache. Often, these complaints are repetitive and stereotyped. It is quite common for people with FTD to have hypochondriacal tendencies. The disease itself does not cause physical pain.