

CFU Carers' Support Group



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

Newsletter

March 2011

Welcome

Welcome to the March edition of the Carers' support group newsletter. Thanks to all of you who attended our meeting on the 21st February. It was our Annual General Meeting (AGM), where we discussed how the meetings have been going and what everyone would like to see in the meetings in the forthcoming year. In addition, we were joined by Dr Anna Richardson, one of our Consultant Neurologists who many of you will know from your clinical appointments, and Professor of Neuropathology David Mann, who runs the Manchester Brain Bank and has been involved in the CFU for many years. They talked to us about recent and ongoing research in frontotemporal dementia, semantic dementia, and progressive aphasia. For those of you who weren't able to attend the meeting we've included a short summary of the key points in this newsletter.

We hope you find the information useful. If you have any unanswered questions or would like some more information on FTD or SD, please 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

If you have any views or ideas on anything discussed at the AGM, please send them to Cheryl Stopford at the Cerebral Function Unit, Hope Hospital, Stott Lane, Salford, M6 8HD (email: cheryl.stopford@manchester.ac.uk). Thank you!

Summary of AGM: 21st February 2011

What do people want from the group? Discussion of future meetings

Meeting times:

- We're very aware that everyone is very busy caring for their loved ones and often may have other responsibilities such as other dependents, full-or part-time work, not to mention all the household duties and trying to fit in some relaxation too! So we're keen to try and accommodate people so they can come along to the meetings but know that it's impossible to please everybody – that's where the newsletter helps to bridge the gap.
- We decided at the meeting to try arranging future meetings for Monday afternoons (starting at around 2-4pm) for a while, trying to avoid traffic and car parking issues at the hospital and to try and enable those who work to make it after an early finish.
- We will also try to develop a basic annual agenda and get the newsletter out in good time so that you can plan your activities around the meetings if you wish.
- If you have any further comments on this, please let us know.

Meeting topics:

- For the next meeting, we have been approached by one of our carers to ask if she could talk to the group about her experiences as a carer. We think that this is a great idea and would be keen to hear from anyone else that would like to share their experiences.
- Some people have noticed that similar symptoms have occurred in several of their family members and want to find out the implications of this for themselves, their children and other relatives. To find out more about this it may be possible to seek help from a 'genetic counsellor'. This is not something that is talked about frequently and people may be unsure of what going to a genetic counselling

session would be like. We thought that it may be helpful to have a session about genetic counselling and what it entails for the second meeting of the year.

- The final meeting of the year is open to suggestions. Please feel free to contact the department!

Group Principles & Ground Rules

Since there are many new members attending the group, and it is a new year, we thought it may be important to specify the 'basic principles' of the group. We also thought it might be useful to set down some ground rules to make sure everyone feels comfortable in the meetings.

Group Principles

The group was started because we realised frontotemporal dementia (FTD) and semantic dementia (SD) are poorly understood in the general community. We felt that this could lead to carers feeling isolated and lacking support.

The focus of the group is to provide diagnosis-specific education and a forum to discuss personally relevant issues.

Proposed ground rules

1. Introductions

We think it is important that at each session we begin by everyone introducing themselves. Since having questions answered is an important aspect of the group, gaining the confidence to introduce yourself to the group may allow you to pose a question to the group later in the meeting. We realise that this may be daunting, but it only has to be short i.e. your name, where you're from, who you are caring for and whether they have FTD or SD.

It may also help you to locate people who live close by to you, caring for someone with a similar problem, who you may want to meet with separately from the group.

2. Confidentiality and anonymity

We ask that what is said in a meeting stays at the meeting. By this we mean not retelling anecdotes you have heard at the meeting to your friends/family and in particular not using peoples' names. Of course it is ok to try out strategies that other carers have suggested that worked for them.

3. Allow everyone to have a say/or not / A place for conversation and a place for silence

The group allows everyone to have an opportunity to share their experiences and stories and to ask questions. However, we realise that it can sometimes be daunting to speak in front of the group and you may not want to, in which case, feel free to just come to sit and listen

4. Allowing everyone a chance to speak

Even in small groups there can be a tendency for some people to talk more than others. Please try to be aware of how much you are saying as we would like everyone to go home from the group feeling like they had an opportunity to speak.

5. A supportive and non-judgemental ear

Listening with an open mind to what others are saying is an essential part of the group. This is not only important when people are sharing their problems, but also when they are trying to provide others/you with advice. For instance, if someone suggests a possible strategy for dealing with an

undesirable behaviour to another member of the group and you say “I tried that and it didn’t work”, it may make the other member of the group feel disheartened, even though that strategy may have worked for them.

6. Focus on the group

During group discussions or talks please try to avoid having side conversations as these can be distracting and cause the group conversation to break up.

7. Avoid interrupting others

When interruptions do occur, try to return the conversation to the person who was speaking beforehand.

8. Realising the impact your words and anecdotes have on others

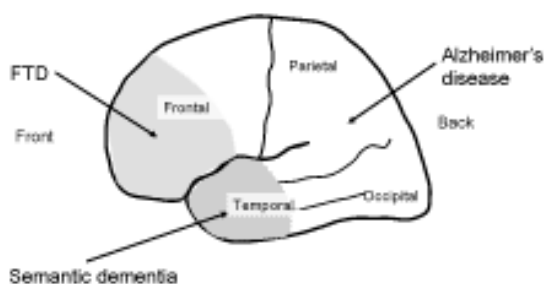
We realise that the group is in some ways ‘cathartic’ and can help you to ‘get things off your chest’ it is also about sharing advice, experiences and information for mutual support. Before sharing an experience, think about how that experience, and the way you recount it, may impact on other people. Focussing on negative experiences can be unhelpful for you and others around you. If you had a problem before, but you got through it, try to focus on how that occurred – what you did to solve the problem and how that has had a positive impact on your life.

Research Activity in the Cerebral Function Unit (CFU)

Dr Anna Richardson and Prof. David Mann

A brief recap of the underlying disease process

Frontotemporal dementia (FTD) and semantic dementia (SD) form part of the same disease, known as ‘*frontotemporal lobar degeneration*’ (FTLD). 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 picture below, in frontotemporal dementia, a disorder of behaviour and personality change, the disease primarily affects the frontal lobes. In semantic dementia, where patients demonstrate impaired understanding and difficulties in communication, the disease selectively affects the temporal lobes. A third disorder, primary ‘progressive aphasia’ (PA), which also affects communication and speech production, selectively affects the left side of the brain. (For more details, see talks from Professor Neary and Dr Richardson on (www.cerebralfunctionunit.co.uk/carers.html))



Research into frontotemporal lobar degeneration (FTLD) is essential to help improve our understanding of the disorders FTLD encompasses, i.e. FTD, SD and PA.

Any research advancements, whether it be in the field of cognition (psychology tests), neuroimaging (brain scans), genetics (blood tests), pathology (investigating the brain at autopsy), pharmacology (medication) or even studies involving animals, can be used together to hopefully find treatments and eventually a cure. The aims of current research are:

1. To improve diagnosis

Many of you will have experienced delays in receiving a diagnosis for your loved one. It is particularly difficult with these types of condition as there are no simple diagnostic tests available. In other areas of medicine, such as dermatology, it is possible to see the problem easily, such as a rash, and diagnose the disease. In addition, these disorders are relatively uncommon, and doctors who aren't specialists are unlikely to recognise them. Therefore, it requires specialist centres like the CFU for accurate diagnoses and, even then, it can still be difficult.

There are different branches to this area of research. For instance, particular psychological/cognitive tests can help aid diagnoses but, as you will all know these tests can be lengthy, can take time to analyse and still may not lead to a clear diagnosis. If a blood test or brain scan could be carried out that was able to provide a definitive diagnosis, it would be ideal, but at the moment this is not the case.

2. To increase understanding of mechanisms/causes of disease

Until we are able to understand what causes a disease we are only able to treat the symptoms, rather than the disease as a whole.

Of particular importance in this area are the genetic and pathological studies.

3. To aid effective treatment

Early and improved diagnosis will be particularly important if treatments are developed: if someone is correctly diagnosed they will be able to receive the correct treatment more promptly.

4. To increase understanding of normal brain functioning

This may not immediately spring to mind as being useful when we want to find out about disorders where people's brains are not working as they should. However, if we do not know how the brain functions normally, it is impossible to work out what and ultimately why something is going wrong in patients with disorders affecting the brain. So, we may ask you, particularly spouses or siblings who are likely to be a similar age to the person who has FTLD, if you would also like to take part in studies.

Neuroimaging

In the department you may have been asked whether you or your loved one might like to take part in a study involving Positron Emission Tomography (PET). PET is a type of brain scan that allows us to find out how well areas of the brain are working i.e. the function of the brain.

You may have heard of MRI and CT scans as these are quite widely used, and there are scanners in most hospitals. MRI and CT scans image the structure of the brain, however, it takes many many brain

cells (neurons) to die before the structure of the brain is altered and for these changes to show up on a structural scan. Therefore, if we rely on MRI or CT scans people have to have had a disease for a long time before any changes in structure can be seen on these scans. In contrast, changes in brain function occur quite early in the disease course and so, by assessing functional changes using PET, we may be able to diagnose patients more easily and at an earlier stage. Unfortunately, PET is not widely used in England and facilities for scanning are sparse. We are lucky to be able to use a PET scanner in Manchester (at the Wolfson Molecular Imaging Centre in Withington) but presently, it can only be used for research, rather than clinically. The current research is therefore very important to improve people's understanding of PET and its usefulness in terms of diagnosis.

Pathology – Understanding cause at a cell and protein level

Even though imaging techniques are always improving, due to the complexities of the brain, some things can only be found out by looking for changes in brain tissue samples. In Manchester we do a lot of work in this area, and we can only carry out these studies if patients/their carers have generously agreed to donate their/their loved one's brain after their death.

So far, we know of three proteins (Tau, TDP-43 and FUS) that accumulate/function abnormally in some patients with FTLD. In some cases these changes in protein function are associated with genetic mutations leading to familial forms of FTLD. In addition to looking at brain tissue, we are also carefully analysing blood samples to look for genes implicated in different types of dementia. This is why you or your loved one may have been asked if you are willing to provide a blood sample when you attended the clinic. If you have not been asked already, it is likely that you will be asked on your next visit to the department!

Although studies have found promising results, so far, these findings do not account for all cases of FTLD. Hence, there is still a great deal of work to do in this area and there are many unanswered questions.

Tau

- In 1998 it was discovered that some patients with familial FTLD, who had accumulation of tau proteins, had a mutation on the gene that encodes microtubule associated protein tau (MAPT) on chromosome 17. This mutation can change the structure of tau, or cause an imbalance of production of tau protein.

TDP-43

- Some people with FTLD have an accumulation of the ubiquitin protein.
- Ubiquitin has a role in clearing waste products from cells.
- Ubiquitin is attached to another cell protein – TDP-43 which has a role in cell nuclei.
- Some people who have an accumulation of ubiquitin have a genetic mutation on the progranulin gene on chromosome 17.

FUS

- This protein has only recently been found to be important in some cases of FTLD.

Questions

People with FTD have often lost their 'common sense'. How can they make the decision to donate their brain?

Since patients with FTD are unlikely to be unable to make a rational decision. You, as someone who knew them before they had the disease, should think about what they would have wanted if they were able to make the decision themselves. This can be difficult as it is unlikely to be something that you would have spoken about. However, knowing the beliefs that the person had can help guide you.

If a patient's brain has been autopsied, have you got the patients' individual records, in particular, details of which proteins were accumulated in their brain?

We know that we have a good correlation between patient diagnosis and their symptoms and the location and type of pathology found at autopsy. However, we do not have information about the individual proteins in each individual, and at present, we would be unable to pass such information on as it would be unethical to do so. We can't be sure about causal relationships or the penetrance of genetic relationships in families as yet, so it wouldn't be appropriate to give advice on these matters that can make a huge emotional impact upon families.

At present, we can only look for these accumulated proteins in brain tissue. But we are looking for markers in blood plasma, this is known as biomarker research and is being carried out in conjunction with increasingly sophisticated imaging techniques and through using blood samples.

It runs in our family, would it be helpful for us (blood relatives) to give blood?

Yes, and we can collect blood when you come with your family member to the clinic.

Are you able to feed back information when patients take part in research?/donate blood samples/brain?

As mentioned earlier, there are ethical issues with this as there are a lot of unanswered questions. However we are currently in the process of getting ethical approval to do this. Even if the information only becomes helpful several years down the line, it would be good to be able to feed back that information at that stage.

How much research has been carried out in the subjects that have been talked about today?

Now, lots! But 10 years ago there was very little. The first gene associated with FTLTD was identified in 1998 and since then there have been huge advances and expansion in the field.

There used to be very little written about dementia in the media, now it seems to be everywhere. Is this because the number of people with dementia has increased or is it because people are living longer?

The number of people with dementia is increasing mainly because people are living longer. Alzheimer's disease, the commonest cause of dementia, becomes more common the older we get. Additionally, compared to previously, diagnosis has improved and dementias are no longer just 'written off' as old age.

However, as you will probably know, FTLTD tends to occur in younger people (aged <65 years).

The three proteins (Tau, TDP-43 and FUS) - do we all have them?

Yes – they all have a normal function. It is only when they do abnormal things, and may not be able to carry out their normal function, that things start to go wrong.

In terms of genetics, does FTLD get passed down to the same sex in families i.e. male-to-male or female-to-female?

There is no evidence of FTLD being sex linked or there being a gender bias. However, in some families there does seem to be a preponderance of one sex with the disorder. This may just be due to chance though.

If it's passed down, does it continue to be passed down?

In some cases it seems to be, whereas in others it doesn't. It is a new area that geneticists are looking into at the moment. However, genetics take a long time to die out so it seems unlikely that it completely stops being passed down.