

MND Podcast number four: Fronto temporal dementia in MND



Hello my name is Robyn Reid. I am the MND Shared Care Worker in the Southern Region of Melbourne. Welcome. This is a series of podcasts on Motor Neurone Disease. This podcast may help you to understand frontotemporal dementia in Motor Neurone Disease. It is thought that MND does not affect people cognitively but in fact fifty percent of people affected by MND will have a mild cognitive impact, while up to fifteen percent will develop frontotemporal dementia. This has a great impact on executive functions of the brain, such as problem solving, decision making, analysis, and personality. Due to the temporal area of the brain being involved, there is also an impact on language. This is in addition to the mechanical difficulties of speech caused by MND. The symptoms of Frontotemporal dementia or (FTD) can be present even before the physical symptoms in the rest of the body. FTD can change a person's capacity to understand and be competent with decision-making which means it is very important to have conversations about end of life wishes as soon as possible. The possibility of being affected by FTD, as well as the problems with speech as the disease progresses, makes these conversations an urgent priority.

The diagnosis of frontotemporal degeneration generally involves:

- Medical history and detailed neurological examination.
- Examination by a Neuropsychologist to assess language, behavior, memory, executive and visual-spatial functions.

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- Neuroimaging to find out where and how severely the brain has been affected. Some of these neuroimaging studies are: MRI (magnetic resonance imaging) and PET (positron emission tomography) scans

Much of the work in diagnosing FTD is in excluding other possible diseases. The only way for certain to know if it is FTD is autopsy.

Given that the cognitive symptoms are sometimes present before the physical symptoms, it is important to check with the carers and loved ones around changes to personality or behaviours. A referral to a Neuropsychologist and Neurologist for an assessment can find out if Frontotemporal dementia is an issue. The multidisciplinary team will monitor for any changes during the course of MND and provide appropriate treatment and support. Carers need to know that the difficulties they are having with their loved one are because of the brain disease, because then they are less likely to feel that their care is not good enough. They may also take up respite, knowing that caring for someone with FTD adds another level of fatigue and distress. It is important to also check that the behaviours of FTD are not creating a lack of safety for the carer e.g., physical, sexual or verbal abuse.

It is important to recognise that FTD can also affect relationships negatively, and quality of life is reduced. Carer burden is greater when there are challenging behaviours as well as physical decline. Some treatments may not be possible due to poor tolerance e.g., pulling out of peg tubes or taking masks off constantly. There may also be changes to people's capacity to understand emotion and comprehend social situations.

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FTD can mean that the person becomes impulsive, loses inhibition or shows inappropriate social behaviour. This may mean that someone who was a polite person, may now become rude or lack tact. They may swear and use language they would never have used before. They may also be physically or verbally aggressive. This can be distressing and embarrassing for carers and loved ones. There is also a lack of insight into their behaviour which makes management difficult. The person may not take into account the needs and feelings of their carers. Emotional changes can include depression, anxiety, paranoia, preoccupation with physical signs and symptoms, and lack of facial expression or detachment from others. The person with FTD may also find it difficult to distinguish between emotions and emotional expressions which may mean they don't communicate well or understand others.

Other impairments can include a personality change, loss of initiative, impaired judgment and poor self-care. These changes are followed by deficits in planning and organization, and inflexible thinking and rigidity.

There can also be other behavioural issues such as repetitiveness. Hyperorality can occur causing changes in eating habits. Hyperorality means people may want the same food all the time or may have inappropriate behavior, such as taking food from others, or demands for sweets. Some people may abuse alcohol, which may be confused as the cause of behavior and not the symptom.

Apathy and adynamia may mean constant monitoring and encouragement to get through simple tasks such as eating or getting dressed. Adynamia means lack of strength or vigor due to an illness. People may also be triggered into

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specific behaviours when they hear certain words and this is called Utilisation. Memory remains intact but there may be an issue with encoding so people may not be able to order or retrieve information correctly. FTD seems to occur more frequently in people with Bulbar and Upper Motor Neurone symptoms.

FTD is not the same as Alzheimers disease. FTD is primarily a disease of behavior and language dysfunction while Alzheimers is one of memory loss. People with FTD do not suffer the same kind of disorientation to time and space and they can keep track of day to day events. Some people with FTD will have language dysfunction which is called Primary Progressive aphasia. This can have identifiable subtypes.

1. Progressive Non-fluent aphasia changes the ability to speak, read and write and understand what others are saying.
2. Semantic dementia creates difficulty finding words and understanding their meaning.
3. Logopenic aphasia makes it difficult for people to perform memory tasks. Speech may be slowed due to trouble retrieving words.

Over time, as things get worse, it is harder for people to understand complex spoken information and there may be additional behavioural features such as irritability, anxiety and agitation. There may also be difficulty walking, rigidity or tremor (similar to Parkinson disease), or muscle atrophy and weakness.

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It is very difficult to look after someone with FTD, so carers will need extra support and more frequent respite. Carers need to develop management strategies for each difficult behavior. A predictable environment, a daily routine and structured activities can help.

Some of the strategies might be to use short feedback on unacceptable behavior and using the same words, so that unacceptable behavior is not reinforced. It is also important to educate friends and family. The person with FTD may have a fixation on a particular food, and without understanding the obsessional way in which the person acts with this food, friends and family who visit may unwittingly bring it to the person. Family and friends also need to know how to manage disinhibition, if that is one of the problems.

People can become aggressive if they are frustrated, therefore anticipating needs and arranging activities around the persons limitations will help. If the person becomes aggressive, defuse the situation by staying calm, don't argue, offer limited choices, set limits, and walk away until the behaviour settles.

Distraction can also be useful. If there are activities that are difficult for the person, such as shopping, try to arrange it so that the person does not need to participate in them.

Where the person lacks initiative and seems to struggle with daily tasks it helps to initiate activity, have everything ready, and prompt if necessary. Offer step by step guidance, and redirect attention to help them refocus. Complete one task at a time, allow sufficient time, and allow for fatigue. Monitor, encourage and reinforce desired behaviours. If certain objects trigger behaviours, remove

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them if possible. If hyperorality is an issue, keep the inappropriate food out of the house and offer something else that can be enjoyed, or limit access.

There are some simple things that can help when dealing with someone with frontotemporal dementia. Set a positive mood for interaction. Your attitude and body language communicate your feelings and thoughts stronger than your words. Set a positive mood by speaking to the person in a pleasant and respectful manner. Use facial expressions, tone of voice and physical touch to help convey your message. Get the person's attention. Limit distractions and noise, turn off the radio or TV, close the curtains or shut the door, or move to quieter surroundings. Before speaking make sure you have their attention; address them by name, introduce yourself by name and role, use non-verbal cues and touch to keep them focussed. If they are seated, get down to their level and maintain eye contact.

State your message clearly. Use simple words and sentences. Speak slowly, distinctly and in a reassuring tone. Refrain from raising your voice higher or louder which they may have difficulty processing, instead pitch your voice lower. If they don't understand, wait a few moments and rephrase the question. Use the names of people and places instead of pronouns and abbreviations. Ask simple answerable questions. Ask one question at a time; those with yes/no responses are best. Don't ask open-ended questions or give too many choices at once. If possible, show the choices as visual prompts and cues to help clarify your question and guide their response. Be patient in waiting for a reply. If they are struggling for an answer it's ok to suggest words.

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Watch for non-verbal cues, and body language, and respond appropriately. Always strive to listen for the meaning and feelings beneath the words. Check with the carer if they have specific gestures that they use for certain words. When the person becomes upset it is important to acknowledge their feelings: “ I can see you are sad - I’m sorry you’re upset - let’s go for a walk.” Distraction, changing the subject or the environment can help to settle the person.

People with dementia are often confused, anxious, and unsure of themselves. Stay focussed on the feelings they are demonstrating and respond with verbal and physical expressions of comfort, support and reassurance as appropriate. Sometimes holding hands, hugging (with permission) and praise will get the person to respond when all else fails.

Carers need encouragement to take time out for their own needs. Establish networks of support that include home-based respite services. Reassure the carer that it may be necessary for them to take over responsibilities and decision-making. Acknowledge the changes in roles and the feelings about it and allow the expression of this.

In summary, Motor Neurone Disease has a spectrum of brain associated symptoms from mild cognitive issues to very complex and hard to manage symptoms with behavior and language involvement, as well as the physical deterioration. The level of brain involvement is on a spectrum from very little to extensive. People with MND may choose to leave their brain and spinal cord tissue for donation upon their death to help with research into MND and

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Frontotemporal dementia. Please contact MNDV for further information about donating tissues. Tissues must be collected within a specific window of time to be viable, so extra care must be taken to communicate that the person is a donor to facilitate their wishes upon their death.

I look forward to discussing more detail about various aspects of MND management in future sessions which will be available on the SMPCC website. I would like to take this opportunity to give my thanks to staff at Bethlehem Calvary health care and MNDVictoria for the support and training received in my role as MND SCW in the Southern Region. Please feel free to contact me should you wish to have an education session on MND for your agency or to receive a secondary consultation. You can contact me on: 0428264446 or email me at: Robyn.reid@smrpcc.org.au

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