Submission to the inquiry by the APPG on Dementia into dementia and comorbidities

Introduction

i. Few conditions are as devastating as motor neurone disease (MND). It is rapidly progressive in the majority of cases, and is always fatal. People with MND will, in varying sequences and combinations, lose the ability to speak, swallow and use their limbs; the most common cause of death is respiratory failure. Most commonly the individual will remain mentally alert as they become trapped within a failing body, although some experience dementia or cognitive change. There are about 5,000 people living with MND in the UK. A third of people with the disease die within a year of diagnosis, and more than half within two years. There is no cure.

ii. The MND Association is the only national organisation supporting people affected by MND in England, Wales and Northern Ireland, with approximately 90 volunteer led branches and 3,000 volunteers. The MND Association's vision is of a world free from MND. Until that time we will do everything we can to enable everyone with MND to receive the best care, achieve the highest quality of life possible and to die with dignity.

iii. As the population as a whole ages, we expect the number of people with MND to increase, and for increasing numbers of people to have both MND and one or more other illnesses, which will include other neurodegenerative conditions such as Alzheimer's. However, this submission will focus on fronto-temporal dementia (FTD), which has significant biological links to MND, which are increasingly becoming understood. Unlike Alzheimer's, FTD does not primarily affect the memory, but rather the ability to take in information.

About MND and dementia

i. Up to 50% of people with MND also experience cognitive change, of whom up to 15% develop FTD. For some people, dementia symptoms occur first: 15% of people diagnosed with FTD go on to develop MND.

ii. Although the presence of a link between the conditions is now well documented, our understanding, continues to develop. Inherited MND (sometimes known as familial MND) accounts for approximately 5-10% of all cases of MND. Mutations in the gene C9orf72 have been shown to be the most common cause of inherited FTD, MND and MND-FTD. Importantly, up to 10% of cases of sporadic MND (where there is no known or clear family history) are also strongly associated with this same gene.

iii. There are three main variants of FTD, which are referred to as:

- behavioural variant (bvFTD)
- progressive non-fluent aphasia (PNFA) or non-fluent variant PPA (nfvPPA)
- semantic dementia (SD) or semantic variant PPA (svPPA).

The variants are not mutually exclusive. Some people show a mixture of problems in language and behaviour. People with MND-FTD will not typically experience all possible symptoms of MND, or all possible symptoms of one of the types of FTD described below. Instead, they tend to present with a combined set of symptoms that are usually from the bvFTD form, along with other language symptoms that could be related to PNFA or SD.

iv. In those with MND affected by cognitive change, the most prominent impairment is in executive functions. These functions are particularly dependent on the frontal lobes of the brain. Some people also show changes in language and behaviour, and some researchers have questioned whether language changes may be at least as common as executive function changes.

v. Memory is not usually affected, although some people may experience problems with their memory as a secondary consequence of executive dysfunction, along with problems with attention. Problems with perception are rarely seen.

vi. Cognitive problems are associated with shorter survival times in MND.

vii. Some people with MND experience emotional lability (also known as pseudobulbar affect). This is not a cognitive symptom, although it can be confused with one by clinicians unfamiliar with it. It can result in uncontrollable laughter or crying in response to something that is only moderately funny or sad, inappropriate responses at embarrassing times, or inability to respond appropriately to other people, which may be interpreted as strange, callous or unfeeling. This can be disturbing to the person themselves and to those around them. Emotional lability is reported to affect 19-49% of people with MND and can occur in those with or without cognitive change.

1. What are the common problems faced by people living with dementia and more than one other health condition? What is your organisation doing to help people overcome these problems?

i. The symptoms of MND-FTD are complex, and the interaction between the physical symptoms of MND and the cognitive and behavioural symptoms of the dementia make managing care for someone with MND-FTD more complex still. Few health and social care professionals fully understand the needs involved.

ii. Care tends to become 24/7 and the person with MND-FTD could have severe behavioural problems as well as issues with capacity. This may include aggression, which creates risk for the carer, other family members and any children in the household.

iii. There is also likely to be inappropriate behaviour, often arising from disinhibition or impulsivity, when people may behave without considering the options and consequences, for instance when crossing the road or handling money; this can even lead to reckless gambling. There can be a tendency to gorge on food, so careful monitoring is required in case the person chokes, as the physical

symptoms of MND often include problems with swallowing – this can mean carers have to remove food from the house or lock drawers and cupboards. The person with MND-FTD often remains undistressed or unaware of the changes they are experiencing, but can become very single-minded and frustrated / angry if something doesn't happen in the way they want it to.

iv. People may also misunderstand how cleaning products and toiletries are to be used, which can pose risks to their wellbeing (for instance eating a dishwasher tablet they believe to be a sweet). Although the sense of taste is not itself affected, people may misunderstand taste sensations, leading to confusions such as adding salt instead of sugar to hot drinks and being unable to distinguish the difference. There are obvious potential dangers associated with this.

v. Onset of MND can occur in any part of the body: if onset is in the limbs, the person tends to experience some immediate loss of independence; if onset occurs with bulbar symptoms however (speech and swallowing), and FTD is also present, the person's independence is less immediately compromised. In this scenario, the dangers already mentioned are heightened, and the person may put themselves at further risk by, for instance, trying to drive or leave the house without their carer knowing.

vi. If the person is tube fed they may reach a point where they no longer comprehend what this is and dislodge or pull out the tubing. Also, if the person is accepted either temporarily or permanently to a dementia ward or residential home, there may be problems associated with tube feeding if not enough nurses are trained to assist. This can place extra burden on carers and family.

vii. Other interventions such as ventilation may need to be risk-assessed if the person with FTD has problems accepting the equipment, or misuses the equipment.

viii. FTD can also cause serious problems for personal relationships. The person may become aggressive or violent without much warning, or appear to be cold and dismissive of others. Disinhibition could cause them to say hurtful things they would not normally and behave inappropriately towards others including in strangers. This could potentially put the person with FTD at risk of abuse or aggression from others who may not understand why they are behaving unconventionally. The person with FTD and their carer can become very isolated as a result.

ix. If speech and communication are physically affected by MND, then FTD will make this situation worse. The person may not be able to voice their needs or be understood, even with communication aids. The APPG on MND received evidence of this during its inquiry into communication support in 2014-15, including the following examples which discuss the use of assistive technology including text-to-speech devices such as Lightwriters and eyegaze systems including software such as GRID.[1]

[1] http://www.mndassociation.org/get-involved/campaigning-influencing/appgreport/

It was a speech typewriter. My Mum used it a little but it was very frustrating for her because she lost the ability to spell, therefore it was very hard to understand what she wanted to say.

At first it seemed OK (Ruth didn't like computers), but she seemed to be giving up, until I read that MND could affect cognitive thought. When I realised that, I deduced that she was struggling to spell the words in the GRID so I got the speech and language therapist to show me the picture version and I programmed that to be suitable for Ruth's needs and she was off and 'speaking'.

As we knew that Jim's speech might be affected we talked a lot early on, but it was so difficult when the FTD put in another barrier – he would write things on the Lightwriter that made no sense, yet it was clear he thought they did. It is hard to say how much this hurt and how difficult my daughter and I found it. Jim was funny and caring - and then he couldn't communicate - we just don't know how this added to his suffering.

A worrying case - a gentleman who had a Lightwriter, but wouldn't use it because he was 100% convinced that his speech was perfectly intelligible, when in fact whatever he tried to say came out as garbled rubbish. He would get very distressed with his family because he thought they were 'having him on' - awful for everyone.

x. The MND Association is taking a range of steps to support people with MND-FTD:
   - Cognition and FTD needs are included in our strategy, which includes requirement to do more to reach affected families
   - We have developed improved information both for professionals and families; cognitive change and FTD are now included in all our resources for professionals, and we are highlighting the availability of the ECAS screening tool (see question 3 below)
   - We raise awareness of FTD and changes to cognition with MND, including at the International Symposium on MND, which we organise
   - We work with experts in the field to better understand the condition, and the latest best practice in relation to screening, assessments and diagnostics.

2) Regarding the management of dementia and co-morbidities, what currently are the chief barriers to improving:

a) **Information provision**

Information tends to be focused on carers, as those with MND-FTD are unlikely to be able to process it themselves. One barrier is therefore getting information to those supporting people affected, as they may be in denial or unaware of why these problems are happening. Reaching and educating the professionals who support these families, who may know little about MND, let alone MND-FTD, is also a challenge.

b) Self-management

The person affected is unlikely to be capable of self-management. Carers and family may misinterpret personality changes – especially where communication

difficulties also exist. Carers and family may hide issues due to perceived stigma, especially when inappropriate behaviour is displayed.

c) **Support**

Adult social care services often do not know how to support people with MND-FTD. Dementia wards and homes often try to deal with MND-FTD like Alzheimer's, but this is often inappropriate. Carers are left exhausted and misunderstood as they try to manage situations on their own. Because only a small proportion of people with MND develop FTD, effective peer is support difficult to find. There is a lack of awareness of the support available in respect of FTD from organisations such as the Alzheimer's Society, Young Dementia and Dementia UK, who offer support for all types of dementia.

3) Regarding dementia and co-morbidities what can the statutory sector (NHS, Local authorities, regulators) do to promote and deliver information, self-management and support?

i. For information, statutory services should refer to the MND Association. Although self-management is extremely difficult for people wit MND-FTD, and often unachievable, more support for carers would be useful. There should also be more specialist care workers trained to manage MND-FTD cases, and clear pathways for the care of people with MND-FTD; we sometimes hear of people being referred back and forth between neurology and mental health services, because responsibility does not clearly rest with either. Clarification of this point in a relevant NHS England specialised service specification may be helpful.

4) Could you outline any areas of best practice in the management of co-morbidities?

i. Formal guidance on good practice is not yet mature. We have developed a booklet for professionals which compiles the best advice on cognitive change and FTD in MND currently available.[2]

ii. Management of people who show signs of cognitive impairment should focus on forward planning and organising appropriate support strategies for them and their families/carers. If cognitive change has been identified, it is important to alert all members of the multidisciplinary team, allowing them to react and implement any changes needed in care planning.

iii. Timely assessment is important. Understanding the level of cognition of the person with MND is crucial to help them and their family to cope with what may lie ahead, including being able to make timely decisions about treatment and care. It is crucial that family members, partners and carers are involved in any assessment of cognitive change, as they will have a perspective on the person's past and present behaviour and personality, and any changes that have occurred. It is important that they are asked to try to identify changes in cognition and behaviour that cannot simply be attributed to the physical symptoms the person is experiencing.

[2] http://www.mndassociation.org/wp-content/uploads/PX018-Cognitive-change-FTD-MND-v1.0-Jun15.pdf

iv. Assessments can lead to suggestions of how to help minimise any confusion and frustration that the changes are presenting. Any management of changes in cognition and/or behaviour should always involve the person with MND, together with their family and carers. Severe cognitive and behaviour change may have implications for adult and child protection issues, so assessment should be prioritised. Such assessments can inform (but not replace) assessments of capacity.

v. The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) tool has been designed specifically as a first step in assessing the presence of cognitive change in MND. The tool is designed to take 20 minutes to complete by any health or social care professional, including non-neuropsychologists. It has been designed to take account of physical disability, as answers can be written or spoken. Part of the assessment includes a carer interview about the behaviour of the person with MND.

vi. Other tools are available. The MiND-B is a simple tool for the identification and quantification of behavioural symptoms in ALS. It measures three behavioural domains: apathy, disinhibition and stereotypical behaviour. The M-ACE tool is a brief and sensitive cognitive screening tool for dementia; unlike the ECAS tool, it is not specific to MND.

vii. Supporting carers is vital. Their individual needs may be complex depending on the severity of cognitive and/or behavioural change in the person with MND. Cognitive and behavioural change is a symptom for which many carers feel unprepared, especially as many people will not have been told that this can occur as part of MND. Clear explanations and instruction can help. The needs of carers and family members should be assessed and support strategies advised.

5) What are the implications for the training and development of the health and care workforce?

i. Inevitably, training on MND-FTD will have to be to some extent specialised. It should probably therefore sit within the specialised multidisciplinary MND clinics nominally commissioned by NHS England but often funded or part-funded by the MND Association under our care centre programme. Training and development will however have to extend beyond these centres to community-based multidisciplinary teams (and looser arrangements) that care for people with MND.

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