FACTSHEET 7

WHAT IS FTD WITH MOTOR NEURONE DISEASE?

Frontotemporal dementia or FTD is a progressive disorder of the brain. It can affect behaviour, language skills and movement.

Motor neurone disease or MND is a disorder of the nerves that control the body's motor function. It affects the nerves that go to the muscles in the arms and legs, and that allow people to speak and swallow.

It is also known as amyotrophic lateral sclerosis or ALS.

About 1 in 10 people with FTD will also develop motor neurone disease. This is known as FTD-MND or FTD-ALS.

It occurs usually in people who have behavioural variant FTD and is less common in people with nfvPPA. It is very rare in people with svPPA.

It occurs more commonly in people who have a mutation in the C9orf72 gene.

MND may present either before or after the symptoms of FTD. In other words, some people who have MND as their first symptom may go on to develop FTD. Similarly, some people who have FTD as their first symptom may go on to develop MND.

In people who have FTD-MND, symptoms may progress more rapidly than in those people with FTD alone, although this is not the case for every person.

Symptoms vary from person to person, but can include:

- Wasting and weakness of muscles
- Twitching of muscles (called fasciculations)
- Stiffness of muscles
- Problems with articulation (production of speech) such that the speech may sound slurred
- Problems with swallowing

Later on in the disease some people develop problems with their breathing.

Does FTD-MND run in families?

FTD-MND can sometimes run in families. Some members of the family may have MND without FTD, or vice versa. This is usually due to a problem in the C9orf72 gene.

See FACTSHEET 2 for more details about familial FTD.

How is FTD-MND diagnosed?

Usually a diagnosis is made by a specialist rather than a GP.

For FTD, there is no single test that will make a diagnosis except in some people who have a genetic cause. A series of tests are usually performed including a scan of the brain.

A diagnosis of MND may be made from the symptoms and signs found on clinical examination. However the diagnosis is usually supported by a series of tests including a needle test of the muscle electrical activity called electromyography or EMG.

See FACTSHEET 11 for more details.

Is there a treatment for FTD-MND?

There is currently no cure for FTD-MND but there are some important things which can help when caring for someone – see FACTSHEET 12 for more details.

People with MND are commonly given a drug called riluzole which has been shown to have some mild benefit. This will usually be started by a specialist in motor neurone disease.

Useful organizations that can provide information about FTD include:

- Rare Dementia Support (UK)
  www.raredementiasupport.org
- Alzheimer's Society (UK)
  www.alzheimers.org.uk
- Association for Frontotemporal Degeneration (US)
  www.theaftd.org
- The Australian FTD Association (Australia)
  www.theaftd.org.au
- For links to other support groups around the world – www.worldftdunited.net

For motor neurone disease, the Motor Neurone Disease Association can provide information and support to people diagnosed with MND – www.mndassociation.org