FACTSHEET 6

WHAT IS NONFLUENT VARIANT PPA?

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

The main subtypes of FTD are called behavioural variant FTD and primary progressive aphasia or PPA. PPA can be further split into three main subtypes, one of which is called nonfluent variant PPA or progressive nonfluent aphasia.

The first symptoms of nfvPPA are usually a subtle change in the ability to produce speech. At first this might be only in certain circumstances such as when speaking on the telephone or when speaking in public.

Symptoms can include:

Slow, hesitant speech

Speech may seem effortful to produce and people may seem to stutter before they can get the right word out. This is known as apraxia of speech.

Difficulty finding the right word to say

Pronouncing words incorrectly

People may say the wrong word, often similar in sound to the one they meant to say, for example 'aminal' instead of 'animal'.

'Telegraphic' speech

People may miss out the small words in sentences such as 'the' or 'and'.

Producing the wrong grammar

People may use the wrong tense such as saying 'happened' when they mean 'happen' – this is known as agrammatism.

Saying the opposite word to the one they mean to say

For example saying 'yes' when they mean 'no'.

Problems with reading

Problems with spelling

Understanding of speech is usually not affected in the early stages.

Non-language functions such as memory also tend not to be affected early on. However, as the disease progresses other thinking problems may occur such as:

- Difficulty understanding speech
- Problems with planning and problem solving

Behavioural problems similar to those seen in behavioural variant FTD may also occur although these often do not occur until the later stages of the disease.

Physical problems occur in some people similar to the symptoms of Parkinson's disease. Very rarely people with PPA may develop motor neurone disease.

It can be a frustrating condition and people may become depressed because of their inability to speak.

Does nfvPPA run in families?

In some cases nfvPPA can be a genetic disorder and run in families. Mutations in the progranulin, C9orf72 or TBK1 genes can cause nfvPPA.

See FACTSHEET 2 for more details about familial FTD.

How is nfvPPA diagnosed?

Usually a diagnosis is made by a specialist rather than a GP. 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. In nfvPPA the scan may show loss of cells in the front of the brain in an area called the frontal lobe.

See FACTSHEET 11 for more details.

Is there a treatment for nfvPPA?

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

The PPA Support Group has been set up to help people with PPA and their carers. It meets a number of times a year. More information can be found at www.raredementiasupport.org