FACTSHEET 4

WHAT IS PRIMARY PROGRESSIVE APHASIA?

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

Primary progressive aphasia or PPA is one of the subtypes of FTD. The three main subtypes of PPA are called:

- semantic variant or svPPA
- nonfluent variant or nfvPPA
- logopenic variant or lvPPA

These are also sometimes called:

- Semantic dementia or SD
- Progressive nonfluent aphasia or PNFA
- Logopenic aphasia or LPA

In all of the types of PPA impairment of language skills is the main problem initially.

Up to 20% of people do not fit into one of the three main subtypes, and are often just called PPA, or sometimes mixed PPA, or PPA-not otherwise specified.

Symptoms of svPPA include:

- Difficulty finding the right word
- Losing understanding of what words mean
- Talking about things in a vague manner
- Difficulty understanding what other people are saying
- Problems with reading
- Problems with spelling

See FACTSHEET 5 for more details.

Symptoms of nfvPPA include:

- Slow, hesitant speech
- Difficulty finding the right word to say
- Pronouncing words incorrectly
- 'Telegraphic' speech
- Producing the wrong grammar
- Saying the opposite word to the one they mean to say
- Problems with reading
- Problems with spelling

See FACTSHEET 6 for more details.

Symptoms of lvPPA include:

- Difficulty finding the right word to say
- Pausing in the middle of sentences
- Difficulty repeating sentences

LvPPA is usually felt to be an unusual form of Alzheimer's disease, and is sometimes called the language variant of Alzheimer's disease.

In all of the different forms of PPA other non-language symptoms occur later on, including:

- Changes in behavior
- Problems with planning and problem solving
- Difficulty remembering things

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

Does PPA run in families?

In some cases PPA can be a genetic disorder and run in families. This is most likely for nfvPPA. Mutations in either the progranulin or C9orf72

genes can cause PPA.

See FACTSHEET 2 for more details about familial FTD.

How is PPA 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.

See FACTSHEET 11 for more details.

Is there a treatment for PPA?

There is currently no cure for PPA 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