

## YOUNGER-ONSET DEMENTIA

### An introduction to younger-onset dementia syndromes

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With advances in nutrition and medicine, longer life expectancies are accompanied by an increase in the number of people affected with dementia. Contrary to common perceptions, dementia is not a single disorder, but rather is an umbrella term used to refer to a number of different clinical presentations, each of which is associated with well-defined changes in the brain. While dementia is typically thought of as affecting older individuals, 5% of individuals under the age of 65 are affected by younger-onset dementia [1]. Here, we discuss the most common forms of younger-onset dementia - Alzheimer's disease and frontotemporal dementia.

#### ***Alzheimer's disease***

Perhaps the hallmark feature associated with Alzheimer's disease is that of memory loss. Certainly, in the early stages of the disease, individuals appear forgetful and seemingly confused when presented with new information [2]. Memories for more recent events are typically disrupted in the early stages of the disease [3].

Disorientation in familiar settings may also occur, leading to problems with way-finding and navigation. Tasks previously undertaken with ease, such as using household appliances, or financial transactions, become increasingly problematic.

The memory problems seen in Alzheimer's disease are the result of progressive damage to brain regions crucial for learning and retaining information. Ultimately, widespread disruption of the brain regions necessary for general orientation, visual and spatial abilities, occurs leading to severe difficulties with everyday functioning.

#### ***Frontotemporal dementia***

In contrast to Alzheimer's disease, the clinical picture of frontotemporal dementia is more variable, with individuals showing changes in behaviour and personality, or changes in language. Three main subtypes of frontotemporal dementia have been established, depending on which abilities are most affected [4].

In **behavioural-variant frontotemporal dementia**, individuals show abnormal behaviours, profound changes in personality, and a reduction in motivation levels [5]. In addition, these individuals may display significant problems in understanding emotional responses in other people, which in turn affects their ability to interact in social situations [6]. Often, the individual will display an inability to inhibit certain behaviours, which may result in moral and social transgressions such as stealing, or making inappropriate comments.

In contrast, individuals with **semantic dementia** experience a progressive loss in their general and conceptual knowledge [7]. These individuals display marked

difficulties in correctly naming objects, recognising musical tunes or familiar faces, and in comprehending the function of certain objects. For example, the individual may refer to a picture of a zebra as a “horse” or a “dog”. Despite their word-finding problems, individuals affected with semantic dementia can converse fluently, and display a range or preserved abilities such as intact everyday and spatial memory.

In contrast to semantic dementia, individuals with **progressive nonfluent aphasia** display profound difficulties with the production of speech. Their ability to communicate becomes effortful with regular pauses in their speech [8]. Using the telephone and communicating with groups of people therefore becomes particularly challenging, and can lead to feelings of isolation in the individual. Importantly, other abilities such as memory, attention, and general comprehension, are not affected. In this regard, computerised technology may be particularly useful to enable these individuals to communicate non-verbally.

### ***Patterns of brain disruption in frontotemporal dementia syndromes***

The contrasting symptoms in each of the frontotemporal dementia variants described above all stem from changes to different regions in the brain. For example in the behavioural variant of frontotemporal dementia, behavioural and social dysfunction is attributable to the degeneration of the frontal lobes. These frontal regions of the brain are known to be responsible for personality, social functioning, and behavioural control. In contrast, in semantic dementia, we see the progressive loss of brain matter in the temporal lobes of the brain. These temporal brain regions are believed to store our general world knowledge, and damage to these regions therefore produces striking difficulties with understanding of the world around us. Finally, in progressive non-fluent aphasia, damage to grey matter tissue deep within the brain separating the frontal and temporal lobes, results in striking communication difficulties.

### ***How is dementia diagnosed?***

A comprehensive investigation is required before a diagnosis of dementia can be made, to exclude any other medical conditions that may be causing the symptoms of concern. Typically these investigations include: detailed medical history, physical and neurological examination, blood and urine tests, brain imaging, and neuropsychological assessment or cognitive testing. Underlying brain pathology, however, can only be determined using advanced neuroimaging methods, or ultimately, at post mortem.

### ***Treatment***

Currently no cure for dementia exists however, management strategies to alleviate some of the cognitive and behavioural difficulties may be helpful. For example, for individuals with memory problems, the use of external aids, such as diaries or setting alarms to remind them of things they have to do, can be useful. For individuals with speech difficulties, using facial expressions, non-verbal gestures, or computerised devices may assist with communication. Dealing with changes in behaviour can be particularly challenging for families and the wider social network of the individual. Accordingly, tailoring the environment to reduce frustration and agitation of the

individual with dementia can help to reduce challenging behaviours. Increasingly, the use of music and art therapy has been shown to improve the overall sense of well-being of individuals with dementia [9]. The next challenge for dementia research lies in the development of genetic testing and new drug therapies which can halt and ultimately cure dementia in the future.

### References:

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### Links to websites for further information:

- FRONTIER, Neuroscience Research Australia  
<http://www.neura.edu.au/FRONTIER>
- Alzheimer's Australia <http://www.fightdementia.org.au/>
- The Association for Frontotemporal Degeneration <http://www.theaftd.org/>