Cognition

Fronto-temporal cognitive changes have been associated with MND. Fronto-temporal dementia is prominent in 5-10% of cases. Recent neuropsychological studies suggest that approximately two thirds of people with motor neurone disease may suffer from mild changes in cognitive skills and processes and/or behavioural change. (MND Australia 2014).

Strong and others 2009

There is considerable evidence supporting the existence of cognitive and behavioural dysfunction in ALS, including a spectrum of frontotemporal syndromes and more classically defined dementias. These include:

- ALS in association with cognitive impairment (ALSci)
- ALS with behavioural impairment (ALSbi)
- ALS with a concurrent dementia that meets the criteria for a FTD (ALS-FTD). The core aspect of this diagnostic categorization relates to the presence or absence of a frontotemporal lobar degeneration. ALS-FTD can manifest in three clinically recognized subtypes.
  - The most common (behavioural variant FTD, bvFTD) is a progressive behavioural syndrome marked by insidious onset, altered social conduct, impaired regulation of interpersonal conduct, emotional blunting and loss of insight.
  - In addition, both a progressive non-fluent aphasia (PNFA – characterized by progressive non-fluent spontaneous speech with agrammatism, paraphasias or anomia) and semantic dementia (SD, characterized by fluent speech with impaired understanding of word meaning and/or object identity) are considered to be within the FTD spectrum.

Miller and others 2009b

What is the prevalence and natural history of cognitive and behavioral impairment in ALS?

- Estimates of cognitive impairment range from 10% (Class III) to 75% (Class III). A population-based sample produced an estimate of 28% (Class II). The prevalence of impairment meeting criteria for dementia ranged from 15% (Class III) to 41% (Class II). Behavioral impairment (irritability and social disinhibition) was identified in 39% (Class III).
- Three studies (Class III) documented mild cognitive decline over 6 months, while others (Class III) found no change over 12 months. It is not known whether patients can progress from ALSci or ALSbi to ALS-FTD. However, 15% of patients presenting with FTD later develop motor neuron degeneration (Class III).
- **Conclusions**
  - A significant proportion of patients with ALS demonstrate cognitive impairment and some have dementia (2 Class II, multiple Class III studies).
- Neither behavioral impairment in ALS nor the natural progression of cognitive or behavioral impairments has been adequately studied.
- **Recommendation**
  - Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation (Level C).