ALS & COGNITIVE CHANGES

FACT SHEET | AMYOTROPHIC LATERAL SCLEROSIS SOCIETY OF CANADA | SOCIÉTÉ CANADIENNE DE LA SCLÉROSE LATÉRALE AMYOTROPHIQUE

Until recently, it was believed that ALS did not affect thinking or behavior. Research in the last few years has shown that ALS can cause cognitive and behavioral changes in some people. Cognitive changes (differences in how people think, know, perceive and understand) usually result from brain cell degeneration in the frontal lobe. Some people with ALS experience behavioral dysfunction without cognitive change. However, many people with ALS willexperience no changes in either thinking or behaviour. The following facts will help you better understand ALS and changes in the brain that may occur. If changes in behavior or thinking are suspected, speak to your ALS medical specialist for further assistance.

- Research suggests that certain proteins, either tau or ubiquitin, may contribute to cognitive impairment
- Chromosomes 17 or 9 may be implicated in familial forms of ALS with cognitive impairment
- Approximately 30 to 50 per cent of people with ALS experience cognitive difficulties
- Mild problems, such as inattention and slowed thinking, are the most common in people with ALS and are often only recognized through neurological testing
- Approximately 20 to 25 per cent acquire severe impairment, such as frontal lobe dementia (FTLD), a progressive condition involving selective degeneration of the frontal and anterior temporal lobes of the brain
- A 2003 study showed approximately five per cent of participants met the full criteria for FTLD, with 52 per cent meeting criteria for possible or probable FTLD
- FTLD appears as a change in personality and/or behavior
- Symptoms include apathy, restlessness, mood swings, loss of reasoning or problem-solving ability, repetitive behaviors
- Unlike Alzheimer's disease, behavioral symptoms are usually the first to develop, rather than memory loss, which may not occur at all
- The most common subtype of FTLD is frontotemporal dementia (FTD), which is marked by early decline in social and personal conduct, lack of emotion and loss of insight
- The two other subtypes are semantic dementia (SD) and primary progressive aphasia (PPA). SD is characterized by loss of vocabulary, poor word comprehension, word finding problems and loss of insight; PA entails non-fluent speech, poor grammar but good comprehension
- People who are older than 60, have bulbar onset ALS, poor breathing or a family history of dementia may be more susceptible to cognitive impairment
- Diagnosis occurs through neuropsychological and neurobehavioral evaluations
- People with both ALS and cognitive impairment may have shorter lifespans than those with ALS alone, possibly because they are less likely to choose life-prolonging procedures due to loss of insight into their ever-changing needs
- Data suggest the pathologic process is somewhat different in people who have both diseases compared with those who have one or the other
- Cognitive problems can't be cured but may be relieved by psychotropic medications
- To accommodate cognitive impairment, family, friends and caregivers can speak slowly and simply, modify the environment and make decisions regarding health-care and finances early in the disease's progression
- Caregivers may need to be involved in the decision-making process when the person with ALS can no longer make appropriate decisions
- Caregivers may require more support when dealing with both ALS and cognitive impairment, as stress and care-giving responsibilities are increased