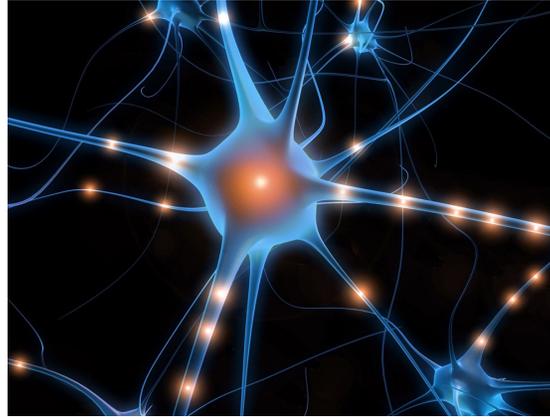


Non-Motor Symptoms in ALS

Cognitive Impairment

Cognitive Impairment in ALS can range from little or no impairment to Frontotemporal Dementia (FTD) associated with ALS.



Pseudobulbar Affect (PBA)

Pseudobulbar affect (PBA) is involuntary and uncontrollable episodes of either laughing or crying that seem inappropriate in the social situation.

Non-motor Symptoms in Amyotrophic Lateral Sclerosis



Cognitive and Behavioral Changes Associated with ALS

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Amyotrophic Lateral Sclerosis Association



Create a world without ALS.

Cognitive Impairment

Prevalence

About 50% of people with ALS develop symptoms of cognitive impairment.

There are 4 subgroups:

- 49% do not develop cognitive impairment
- 45% develop mild to moderate changes
- 6% develop FTD

Mild Cognitive Impairment in ALS

These mild symptoms might be so subtle that they are only detectable by neuropsychological testing or they may cause slight frustration for the patient.

These symptoms may include:

- Poor planning
- Difficulty with problem solving
- Inattention or distractibility
- Poor word generation
- Short-term memory problems
- Language difficulties

Implications

The presence of cognitive impairment may call in to question the ability of ALS patients to make medical and end-of-life decisions.

Neuropsychological evaluations may be beneficial in helping the family make future plans.

Medical and financial powers of attorney should be discussed to verify the patient's wishes are followed.



FTD in ALS

If changes in cognition and behavior begin about the same time as the motor symptoms, are severe, and interfere with the person's ability to perform their daily routines, it is diagnosed as FTD associated with ALS.

FTD is often associated with brain shrinkage in the frontal and temporal lobes of the brain. There are no lab tests that can be done. The diagnosis is made through interviewing the patient and family about the symptoms and family history and through neuropsychological testing.

FTD Behavioral changes include:

- Lack of interest or withdrawal
- Reduced spontaneity
- Lack of emotion or an increase in emotional reactivity
- Loss of inhibition or inappropriate social behavior
- Changes in sleep and eating patterns
- Repetitive behaviors such as hoarding or craving foods

FTD Cognitive changes include:

- Inattention
- Poor planning or organization skills
- Memory loss
- Loss of language
- Problem solving difficulties or poor judgment

Treatment of FTD in ALS patients

There is no specific treatment. Doctors focus on trying to lessen the symptoms and help the family cope with the changes.

- Rilutek, Neurontin, and Radicava, medications often used to treat ALS, have not been shown to improve cognitive impairments in ALS.
- Aricept, Exelon, and Reminyl, commonly used medications to treat Alzheimer's disease, have been shown to worsen irritability in FTD.
- SSRIs, such as Prozac, Zoloft, and Celexa and dopamine blockers, are sometimes used to treat behavioral disturbances in FTD.

Pseudobulbar Affect (PBA)

Pseudobulbar affect (PBA) is involuntary and uncontrollable episodes of either laughing or crying that seem inappropriate in the social situation. PBA can cause a great deal of distress in social situations, in the work place, and with family.

The emotion is often triggered by something only slightly funny or sad. For example, a sad TV commercial may trigger uncontrollable crying.

The emotion may also be the opposite emotion that the person is feeling. For example, a fit of uncontrollable laughter may be triggered by feelings of intense anger.

Prevalence

Between 43% and 49% of individuals with ALS exhibit PBA. PBA is also found in individuals with stroke, traumatic brain injury, multiple sclerosis, Parkinson's disease, and Alzheimer's disease.

Treatment Options

Pharmacological:

Nuedexta is currently the only pharmacological treatment approved by the FDA for PBA.

Antidepressants, such as amitriptyline, fluoxetine, and citalopram, can also be used to treat PBA.

Behavioral Strategies:

Eliciting the opposite emotion can help control their emotions. For example, during an episode of uncontrollable crying having someone tell a joke can stop the crying spell.

