Cognitive and Behavioral Changes in ALS: 
A Guide for People with ALS and their Families

Overview

If you had asked doctors about ALS 15 years ago, the majority of providers would have told you that ALS only affects the motor and breathing functions of the body but that the mind remains normal. Thinking and behavior changes were observed in individuals with ALS and documented in scientific and clinical papers back in the 1800s. Despite these reports, though, early research and clinical care in ALS was importantly focused on establishing the diagnosis of the disease, identifying and testing techniques allowing early diagnosis of ALS physical symptoms, and the development of clinical trials and programs allowing individuals to live higher qualities of life with the progressive physical challenges of the disease.

Within the last 15 years, and within the last 10 years particularly, thinking and behavior symptoms are increasingly recognized in people with ALS. Our knowledge is rapidly growing about how thinking and behavior are affected in ALS. Specifically, we are all learning how thinking and behavior changes impact disease course, symptom management, and decision making throughout the illness. Genetic discovery, brain imaging studies, and biomarker studies now provide tangible explanations for why changes in thinking and in behavior can be symptoms of ALS. Research on the highly variable presentation of symptoms in ALS has grown significantly and further work is underway to address how to predict who will develop thinking and behavior symptoms and how we can improve care and clinical trials for individuals with those symptoms.

The presentation of ALS varies from person to person. Some people with ALS will never develop changes in thinking or behavior. For others with ALS, there will be mild changes in how they think or behave but they are still able to function independently and make informed decisions about their care. Finally, for some people with ALS, changes in thinking and behavior are quite significant and severe such that these people are challenged to make informed decisions about their care and activities and require others to act on their behalf. Educating people about thinking and behavior changes unique to ALS helps to empower a person with ALS, to validate the experience of caregivers and family members, and to educate providers working with an affected person so that decisions are made in a manner consistent with honoring the individual’s longstanding values, preferences, and desires.

How many people with ALS experiencing changes in thinking and behavior?

Current research data suggest that up to 50% of people with ALS will never develop significant changes in thinking or behavior, over and beyond normal psychological reaction to diagnosis and symptoms. That said up to 50% of people with ALS will experience some degree of change in thinking or behavior, with approximately 25% of those people with ALS developing a full blown dementia.
What is the difference between “impairment” and “dementia”?

Healthcare providers may refer to “impairment” or “dementia” when discussing thinking and behavior symptoms. “Impairment” recognizes that the person with ALS is acting in a way that is different than who he/she has always been but not to the extent that he/she cannot still complete activities and think through decisions as he/she has always done. “Dementia” recognizes that the person with ALS is acting in a way that is different than who he/she has always been AND he/she can no longer complete activities and think through decisions as he/she has always done. Different diseases can cause dementia. We now know that ALS can, but does not always, result in dementia. The type of thinking and behavioral impairment observed in ALS is often different than the rapid forgetting that marks the onset of Alzheimer’s type dementia.

What do thinking and behavior changes in ALS look like?

Different symptoms can develop in different individuals with ALS. Here are signs and symptoms commonly seen when behavior and/or thinking are impaired in ALS:

- Behavior becomes embarrassing, childlike, inappropriate, or uncharacteristic
- Person seems to have lost “a filter” with regard to making comments or expressing opinions
- Person begins eating sweets, or only one type of food to the exclusion of a more balanced diet
- Person loses table manners and begins stuffing their mouth with food
- Decreased attention to hygiene such as toileting, bathing, grooming, or changing clothes on a regular basis
- Loss of judgment with regard to making decisions or making a decision that reflects a strong departure from views the person expressed in the past
- Lack of concern for others, one’s own illness and symptoms, and/or no view of the future
- Inability to concentrate or to shift focus from one activity to another
- Fixation on a single idea or activity with a need to repeat the concern or repeat the activity
- Increased aggression
- Says “no” when means “yes,” or becomes less reliable with yes/no responses
- Feels like there is a disconnect between having the thought to move and being able to move the intended body part
- Writes or says words in the wrong order or without respect to grammar
- Thinks of the word he/she wants to use but cannot get it out in conversation
- Loss of spelling or loss of word meaning
- Says sentences that convey little meaning
- Cannot follow instructions to complete physical therapy/occupational therapy/speech therapy exercises, stretches, or guidelines such as swallowing precautions
- Difficulty remembering what he/she intends to do

Impairment in thinking and behavior within ALS can be due to the manifestation of the disease but can also relate to other factors such as insufficient breathing mechanics (too little oxygen or too much carbon dioxide), medication side effects, depression or anxiety, sleep disturbance, or pre-existing
psychiatric or neurologic disease. It is important to bring any symptoms to the medical providers so that providers can begin to evaluate the cause of the impairment, particularly if there is any chance that it can be reversed.

**Risk Factors for Cognitive and Behavioral Impairment in ALS**

Although older age, bulbar onset disease, family history of dementia, and pre-ALS neurologic injury have been cited as risk factors for developing cognitive and behavioral impairment in ALS, there are examples of individuals who develop the symptoms without these risk factors. Currently the only consistently documented risk factor for the evolution of cognitive or behavioral impairment in ALS is the presence of abnormal repeats in a gene called “*C9ORF72*.”

**Diagnosis of Cognitive and Behavioral Impairment in ALS**

In order to evaluate whether the cognitive and behavioral symptoms you see are related to ALS or another process, you will be referred to a neuropsychologist for cognitive evaluation. The neuropsychologist will give you various paper and pencil tests to determine how you process information. You may be asked to recite as many words as you can, beginning with a specific letter of the alphabet. You may be asked to remember words or stories and say these back to the examiner. You may be asked to spell some words. The data collected are compared to normative data for people of your same age and education and compared to estimates of your longstanding level of function. This helps to determine if there is impairment and what type it could be. The neuropsychologist may speak to the person with ALS and also a caregiver or family member who knows the person well.

**What happens after cognitive or behavioral impairment has been diagnosed?**

If impairment is diagnosed, this may come as a relief to family or caregivers. Often times when a person has cognitive and behavioral changes in ALS, it can be a relief to know that the person is not acting abnormally due to psychological reasons or trying to be oppositional or challenging to others. If insight and self-awareness is present, an impaired person can make sure that he/she attends to advanced directives and documents wishes and intentions for later on in the disease process. This is particularly important because cognitive and behavioral impairments, like other symptoms of ALS, advance with disease progression and worsen over time.

If insight and self-awareness are absent, it becomes very important for family members, caregivers, and even health providers to set realistic expectations for the person. Without insight, a person cannot be expected to change his/her behavior. The environment around the impaired person must change. Expectations for the person should match the person’s ability level. If the cognitively/behaviorally impaired person starts to withdraw or starts to become agitated in the face of requests for certain action, these can be signs that the expectations of the situation exceed ability. Likewise, if a family member, caregiver, or care provider starts to experience more frustration or irritation in working with the impaired individual, then expectations of the situation may have to be simplified. In the context of dementia, speech therapists, occupational therapists, physical therapists, neurologists, and others...
working with the affected person should direct all education and interventions towards the caregivers and family as the affected person will not and cannot be expected to change on his/her own.

Family members or caregivers dealing with frustration in the context of cognitive and behavioral impairment in ALS may benefit from supportive counseling, support groups, spiritual counseling, or increasing their own focus on taking care of themselves so that they have more resources (physical and emotional) to offer the person with ALS.

**How do these additional symptoms impact ALS disease course and treatment?**

There are data showing that people with cognitive and/or behavioral impairment in ALS live a shorter life span than people without such impairment in ALS. Many studies are actively in process to determine if the presence versus absence of cognitive/behavioral impairment impacts the effectiveness of medications, treatments, or other procedures in ALS management. This is an important area of research.

**A word about pseudobulbar affect:**

Some people with ALS develop an unusual symptom called “pseudobulbar affect.” They may cry or laugh at inappropriate times or discuss how once they start feeling an emotion, it is difficult to shut it off. Sometimes, pseudobulbar affect can be present when a person feels more emotionally reactive in general, with more intensity to the emotion that is experienced than normal. Pseudobulbar affect is common in ALS and is the result of a brain reflex that is no longer working correctly. It does not necessarily mean that a person is feeling anxious, sad, depressed, or emotionally distraught. People with ALS can have pseudobulbar affect and no other cognitive, behavioral, or psychological symptoms.

**Recommendations for care and support of someone with ALS and cognitive or behavioral impairment:**

- Educate yourself.
- Take care of yourself.
- Simplify communication with the affected person. Break sentences up into short phrases. Ask yes/no questions. Slow down when speaking.
- Provide supervision and accompany the person to all appointments to make sure information is accurately relayed and retained.
- Set realistic expectations for the person with ALS. If your requests introduce frustration, irritability, or withdrawal for either you or the person with ALS, your expectations need to be modified and likely simplified to meet the needs of the person’s current thinking abilities.
- Educate providers and caregivers working with the affected individual about where to set expectations for the affected person.
- Continue to enjoy relationships that bring joy and take breaks from relationships that add to stress.
- Continue to enjoy activities that bring joy and can be conducted safely. Refrain from activities that result in stress or result in risk of safety or liability.
The ALS Association thanks and acknowledges Beth K. Rush, Ph.D., ABPP from the Mayo Clinic for sharing her time and expertise on revising this factsheet. In addition, The Association thanks J. Murphy and M. Lyon for their contributions to this FYI.

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Revised October 2014 by Beth K. Rush, Ph.D., ABPP.

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