

Changes in Thinking and Behavior in ALS



LIVING WITH ALS
RESOURCE GUIDE



Changes in Thinking and Behavior in ALS

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A NOTE TO THE READER: The ALS Association has developed the *Living with ALS* resource guides for informational and educational purposes only. The information contained in these guides is not intended to replace personalized medical assessment and management of ALS. Your doctor and other qualified health care providers must be consulted before beginning any treatment.



Introduction

Twenty years ago, most would have told you that ALS only affects the motor and breathing functions of the body; the mind remains normal and unaffected. Yet thinking and behavior changes were observed in people with ALS and documented in scientific and clinical papers back in the 1800s.

Within the last 20 years, it has become clear that thinking and behavior symptoms are common to the experience of ALS. We now know that when thinking and behavior changes evolve in ALS, these symptoms significantly impact disease course, symptom management and decision making throughout the illness. Genetic discovery, brain imaging studies and biomarker studies now provide tangible explanations for the presence of behavior and thought change in ALS.

The following topics are covered in this resource guide:

- The role of the brain in thinking and feeling
- The connection between ALS and changes in thinking and behavior
- The frequency of thinking and behavior changes in ALS
- The difference between “cognitive impairment” and “dementia”
- What changes in thinking and behavior look like in ALS
- Risk factors
- Diagnosis and what happens after
- How cognitive and behavioral changes impact the course of the disease and treatment
- Pseudobulbar affect disorder

Your Brain as a Thinking and Feeling Network

Our brains control many functions of the body, including our thoughts, our feelings and our reactions. Different areas of the brain have specific functions when it comes to thinking.

The brain has four major lobes:

1. Frontal lobe
2. Parietal lobe
3. Temporal lobe
4. Occipital lobe

These brain areas have cell bodies, or neurons, that are designed to process specific types of information. For example,

the temporal lobes of the brain help us take in and keep memories and learn associations between words and experiences. The frontal lobes of the brain help us coordinate movements, speak, organize, plan and control our urges, such as laughter, anger, fear, hunger and thirst.

In addition to these four major systems for thinking and behavior, we have long channels of communication between neurons in each lobe, between left and right sides, and from top to bottom (and vice versa) in the brain. These long channels are made up of **axons** or white matter. If there is enough change in neurons, axon tracts connecting functional areas of the brain become disconnected.

Any potential injury to the brain—damage to neurons or axons—can result in a change in how we think and how we behave.

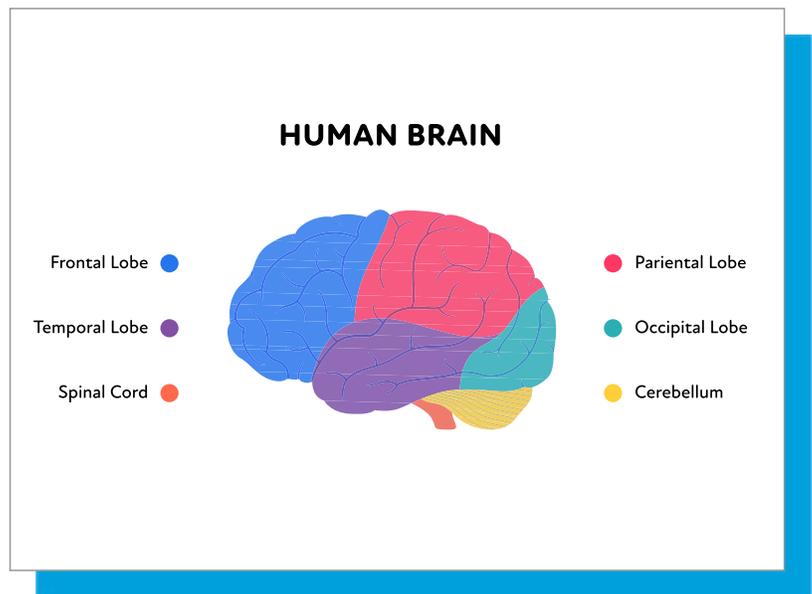


Figure 1: Areas of the Brain

Sometimes that injury is very specific and limited to one place and one function, such as being able to find our words in conversation. Other times, the injury disconnects one part of the brain from communicating with another important part of the brain. For example, if disease impacts frontal lobe connections, a person can lose insight into how they have changed or how behavior impacts others. This can happen even if there is no injury to the frontal lobe itself. Our brains are a very sophisticated network of signals, chemicals, and structure that controls behavior.

What Does ALS Have to do with Thinking and Behavior?

A person with ALS may have symptoms of disease outside of the motor areas in the brain. In particular, the frontal and temporal lobe systems can be impacted. When this area of the brain is

affected, there will be changes in thinking and/or behavior. Some studies have shown us that the cerebellum can also be affected.

For some people, changes in thinking and/or behavior can show up well before any motor symptoms. For others, changes in thinking and behavior come after motor symptoms have already started. Finally, it is possible for changes in thinking and behavior to occur at the same time as the motor symptoms. It depends on where the disease starts in the brain and how it spreads. This is why different people with ALS have such different journeys. You may pick up on this if you attend a support group and listen to people discuss how ALS has impacted them differently.

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How Many People with ALS Experience 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.

Up to 50% of people with ALS might experience some degree of change in thinking or behavior, with approximately 20% of those developing a form of dementia called frontotemporal dementia.

At this time, we do not understand the science behind why certain people experience certain symptoms. We also do not know if there are protective factors, such as keeping busy, continuing to learn or doing puzzles, will have any impact on how disease spreads and symptoms develop. Further work in this area is being done to help answer these questions.

What is the Difference Between “Cognitive Impairment” and “Dementia”?

Healthcare providers may refer to cognitive “impairment” or “dementia” when discussing thinking and behavior symptoms. When there is ALS with cognitive impairment or behavioral impairment, you may see it written as ALS*ci* or ALS*bi*, respectively.

“Impairment” is when a person with ALS is acting in a way that is different than who they have always been but **not**

to the extent that they cannot still complete activities and think through decisions as they have always done.

“Dementia” is when the person with ALS is **acting in a way that is so different than who they have always been AND they can no longer complete activities and think through decisions** as they have always done. For a person to meet criteria for dementia, it is important that providers and family consider what the person with ALS could do for themselves if motor limitations (e.g., loss of speech, loss of limb movement or ambulation) were not present. Different primary neurodegenerative diseases cause dementias. ALS is not typically a memory loss dementia such as that which we see in the dementia associated with underlying Alzheimer’s disease. However, there are some most rare cases of ALS for which memory is impacted.

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
- **Think of the word they want 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 they intend to do**



There can be what we call "secondary" causes of cognitive and behavioral change in ALS. For example, people with ALS may struggle with breathing mechanics (resulting in too little oxygen to the brain or too much carbon dioxide in the body). There

are medications that are used to manage motor symptoms of ALS that have side effects. Depression, anxiety and sleep disturbances can occur with adjustment to ALS, and these factors can have some degree of impact on thinking and behavior. 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.

Older age, bulbar onset disease, family history of dementia and pre-ALS neurologic injury may put a person at higher risk for developing cognitive and behavioral impairment in ALS. However, there are examples of individuals who develop the symptoms without these risk factors.

Risk Factors for Cognitive and Behavioral Impairment in ALS

Older age, bulbar onset disease, family history of dementia and pre-ALS neurologic injury may put a person at higher risk for developing cognitive and behavioral impairment in ALS. However, 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. If speaking your answers is not possible, the neuropsychologist should give you an opportunity to write your responses. If writing is not possible for you, the neuropsychologist should give you the opportunity to speak your answers or to nod "yes/no" to questions you are asked.

The data collected are compared to how people of your same age and education without impairment perform 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 a caregiver or family member who knows the person well. Evaluating thinking and behavior in ALS can be helpful for everyone to understand how the disease is expressing itself and to prioritize decisions that need to be made about managing symptoms that come along with living with ALS.



Why It's Important to Know Changes in Thinking and Behavior May Be Part of the ALS Journey

Since we have developed greater awareness of thinking and behavior symptoms in ALS, a very common question asked is, "Why would I want to know if impaired thinking or behavior is part of my ALS journey?"

Research on ALS as a disease is making discoveries at a rapid rate. The last 20 years are a true testament to this.

One of the most important focuses in the context of any progressive illness or disease is quality of life. With ALS, there are many factors you will not be able to control, but how you make decisions, what your values and beliefs are and how you want to be treated should be very much within your control. In a study that polled people with ALS about whether they would want to know if thinking or behavior impairment was present, a majority of people responded, "Yes!"

- **Being able to know if you have impairment or not is helpful in decision-making regarding symptom management.** If you have no changes in thinking and behavior, you continue to make decisions about care, nutrition, support, relationships and therapies available to you.
- **If you have any impairments in thinking or behavior, then it becomes very important to identify a trusted person who can act on your behalf,** taking all information into consideration. This person can balance information presented about your symptoms and disease management with your longstanding values and beliefs, honoring you in a way that you may not be able to do for yourself.

Research on ALS as a disease is making discoveries at a rapid rate. The last 20 years are a true testament to this. Your doctors

are very interested in developing new treatments for ALS. The presence of ALS with symptoms of thinking and behavior may make you eligible for different treatments. Further, **the presence of these non-motor symptoms may make it such that you respond to known treatments and medications differently than a person with ALS who does not have these non-motor symptoms.**

What Happens After Thinking or Behavioral Impairment Has Been Diagnosed?

When thinking and behavior impairments are diagnosed, many family members and caregivers feel a sense of relief. Often family members, in particular, notice differences in a person's thinking or behavior but believe it's just a part of how the person is adjusting to the disease or coping with the disease. It relieves some pressure to know that others can see changes in the person with ALS and that it is not simply something happening in your relationship.

When you are a family member observing the changes in someone with ALS you may be concerned that commenting on the change may be judging the person with ALS rather than helping the situation. **In some situations, when a person has thinking/behavior changes in ALS, it can be helpful to know that the person is not trying to be oppositional, argumentative, childlike or challenging to others.**

It can be particularly helpful to know—in cases when a person has a frontal lobe injury, or network disconnection, from disease—that **poor insight and self-awareness of symptoms and behavior are part of the disease rather than a psychological reaction.** Knowing whether a person with ALS has thinking/behavior change helps those who interact with the person know when to allow the person to act for themselves versus responding and acting on their behalf.

When insight and self-awareness remain present in a person with ALS, the person can make sure that they attend to advanced

directives and document wishes and intentions for later 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 their behavior. The environment around the impaired person must change. Expectations for the person should match the person's ability level.

If a person with thinking/behavior impairment 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 healthcare provider starts to experience more frustration or irritation in working with the impaired individual, 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 toward the caregivers and family as the affected person will not and cannot be expected to change on their own.

Family members or caregivers dealing with frustration in the context of thinking/behavior impairment in ALS may benefit from supportive counseling, support groups, spiritual counseling or increasing their own self-care so they have more resources (physical and emotional) to offer the person with ALS.

How Do Thinking and Behavior Symptoms Impact ALS Disease Course and Treatment?

Some studies have shown that people with thinking/behavior impairment in ALS live a shorter life span than people without such

impairment. Many studies are actively in process to determine if the presence versus absence of this 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 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 Caregivers

- ✓ **Educate yourself.** Read everything you can about ALS. Information about the disease and treatments available changes rapidly as research updates how doctors and therapy providers practice. Attend support groups if you are comfortable doing so. Speak to ALS Association staff and your clinical care team about any concerns you have and to discuss further evaluation of the thinking/behavior changes you see.
- ✓ **Take care of yourself.** Remember what our flight attendants tell us just before every flight we take: "Make sure to secure your own oxygen mask before assisting others." What does this mean in caring for someone with ALS? When you are a caregiver, make sure you are sleeping regularly, eating well and managing stress.
- ✓ **Keep engaged in relationships and activities that bring you joy.** Limit those that result in stress. If you are feeling overwhelmed, ask yourself, "What can I do to make myself feel better right now? What can I do to make myself feel better over the next month?" Do not neglect your own regularly scheduled appointments and activities. You need to make sure you give enough to yourself and budget withdrawals of your energy so that you have something to give the person with ALS you are trying to support.
- ✓ **Simplify communication with the affected person.** Break sentences up into short phrases. Ask yes/no questions. Slow down when speaking. If a person cannot speak, write down two choices. Have the person point to a response or give an eye gaze to the selected response. Slow down.
- ✓ **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.

- √ **Set realistic expectations for yourself.** If you are feeling overwhelmed, it is time to consider what you have committed yourself to and whether you can do it. People are not superheroes. Even when you love someone tremendously and want to demonstrate commitment, you have to take care of yourself and acknowledge realistic limitations. Do not wait until you are feeling "under water" in trying to manage care. Think ahead and talk with your local care team and ALS Association staff about what options you have for help and/or time away, if required.

- √ **Educate providers and caregivers working with the person who has thinking/behavior impairment about where to set expectations for the person.** In an ideal world, all healthcare providers would be on the same page about what to do and how to accomplish it. That said, many healthcare providers may not have much experience with ALS and how it affects people. You may need to share the knowledge you have learned through The ALS Association and your own understanding of the disease to stand up for the person with ALS and get the services and care required.

- √ **Continue to enjoy activities that bring joy and can be conducted safely. Refrain from activities that result in stress or risk of safety or liability.**

Summary Statement

Prior to 20 years ago, most people working in the ALS field would say that ALS does not affect thinking or feeling. That has changed.

Not everyone diagnosed with ALS will experience changes in thinking and behavior. It is assumed that at least 50% will, and the changes can range from mild to severe in nature. Knowing that this is potentially part of the ALS disease is useful because if changes are noted, you know it is not just an emotional response to coping with the diagnosis, but a part of the disease process. This has an impact on decision making. Choices about treatments need to be addressed early on so a person's wishes are known and a trusted loved one can ensure decisions are honored. Also, additional support for caregivers who may be challenged by changes in their loved one's thinking and behavior may need to be arranged.

Further research is being done to help understand why the parts of the brain involved in thinking and behavior are affected in some people and not others, risk factors, prognosis and what treatments may be more effective than others when there are changes in cognition.



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About The ALS Association

The ALS Association is the largest philanthropic funder of ALS research in the world. The Association funds global research collaborations, assists people with ALS and their families through its nationwide network of care and certified clinical care centers, and advocates for better public policies for people with ALS. The ALS Association is working to make ALS a livable disease while urgently searching for new treatments and a cure.