



Changes in Thinking and Behavior in ALS



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.

Living with ALS
Changes in Thinking and Behavior in ALS

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INTRODUCTION

If you had asked doctors about ALS 15 years ago, most would have told you that ALS only affects the ability to move the muscles in your body (medical term: **motor**), but that the mind remains normal.

Within the past 15 years, and within the past 10 years in particular, thinking and behavior symptoms are increasingly recognized in people with ALS. These symptoms are also called **non-motor symptoms**. Our knowledge is rapidly growing about how thinking and behavior are affected in ALS. Specifically, we are learning more details about 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. 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 people with those symptoms.

What we will cover in this resource guide:

- The role of the brain in thinking and feeling
- The connection between ALS and changes in thinking and behavior
- How common changes in thinking and behavior occur
- 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 (PBA, also known as pathological laughing and crying)

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 and each has specific functions (**Figure 1**):

1. Frontal lobe – Speaking, planning, behavior, and movement
2. Parietal lobe – Processing touch and sensations
3. Temporal lobe – Hearing, understanding the meaning of words, learning, emotions, and memory
4. Occipital lobe – Sight

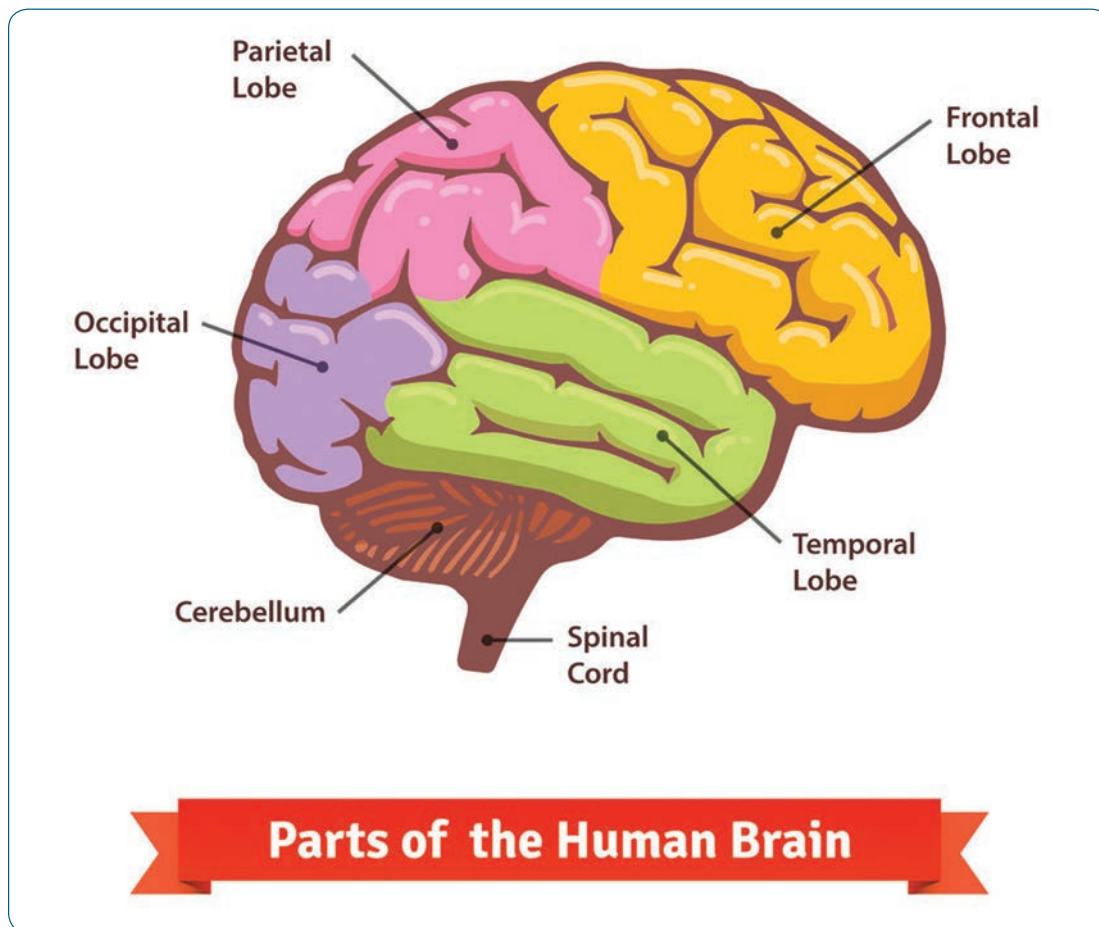


Figure 1: Areas of the brain.

These brain areas have nerve cell bodies, or neurons, that are designed to process specific types of information. For example, the temporal lobes of the brain help us to take in and keep memories and also help us learn associations between words and experiences. The frontal lobes of the brain help us to coordinate movements, speak, organize, plan, and to control our urges such as laughter, anger, fear, hunger, and thirst.

There are channels of communication between neurons in each lobe that make connections between left and right sides, and from top to bottom (and vice versa) in the brain. These long channels are made up of **axons**. **Any potential injury to the brain, damage to neurons or axons, can result in a change in how we think and how we behave.** Our brains are a very sophisticated network of signals, chemicals, and structure that controls behavior. Sometimes an injury is very specific and limited to one place and one brain 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 the connections of the frontal lobe, a person can lose insight into how he/she has changed or how behavior impacts others. This can happen even if there is no injury to the frontal lobe itself.

ALS AND THINKING AND BEHAVIOR

The brain of a person with ALS can be affected outside the areas that control movement. In particular, the frontal and temporal lobe systems can be impacted. When these areas of the brain are affected, there will be changes in thinking and/or behavior.

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

Number of People with ALS Who 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 a normal psychological reaction to diagnosis and symptoms.

That said, **up to 50% of people with ALS might experience some degree of change in thinking or behavior, with approximately 15% of those developing 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, that will have any impact on how disease spreads and symptoms develop. Further research in this area is being done to help answer these questions.

COGNITIVE IMPAIRMENT AND DEMENTIA

Healthcare providers may refer to cognitive “impairment” or “dementia” when discussing thinking and behavior symptoms.

“**Impairment**” is when a 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**” is when the person with ALS is **acting in a way that is so 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. Memory loss does not need to be present to have dementia. Many diseases other than ALS can cause dementia.

We now know that ALS can, but rarely results in dementia. The type of thinking and behavior impairment observed in ALS is often different than the rapid memory loss that marks the onset of Alzheimer’s type dementia.

Thinking and Behavior Changes in ALS

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
- **Shows decreased attention to hygiene** such as toileting, bathing, grooming, or changing clothes on a regular basis
- **Losses of judgment** with regard to making decisions or makes decisions that reflect a strong departure from views the person expressed in the past
- **Lacks concern for others**, one’s own illness and symptoms, and/or has no view of the future
- **Is unable to concentrate** or to shift focus from one activity to another
- **Fixates on a single idea or activity** with a need to repeat the concern or repeat the activity
- Shows increased aggression
- **Becomes less reliable with yes/no responses** or says “no” when means “yes”
- **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
- Loses the ability to spell or cannot understand the meaning of words
- 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
- Shows difficulty remembering what he/she intends to do

Cognitive impairment in ALS can be due to the disease itself, 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 disturbances, 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

Older age, ALS that starts in the speech or swallowing muscles (medical term: **bulbar onset disease**), a family history of dementia, and neurologic injury before the onset of ALS 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.

Research has discovered a gene named C9ORF72 that increases the risk for having changes in thinking and behavior in people with ALS. Testing for this gene is available and something you can discuss with your healthcare provider.

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 **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 you have difficulty speaking, you may be able to write your responses. If you have difficulty writing, you may be able to speak your responses.

The information collected is 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 as well as to 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.

Knowing That 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?"

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 how to go about 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 impairment 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 long-standing 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 revealing discoveries at a rapid rate. The past 10 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 changes in addition to weakness may make you eligible for different treatments. Further, **the presence of these symptoms may make it such that you respond to known treatments and medications differently than a person with ALS that does not have these symptoms.**

After Thinking or Behavioral Impairment Have Been Diagnosed

I am so relieved to know this information. I thought I was doing something wrong. Now I know it is no one's fault.

Anonymous Caregiver (Contributed by The ALS Association Golden West Chapter)

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 is 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 or bringing it to light 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 caregivers to know that the person is not trying to be oppositional, argumentative, childlike, or challenging to others.**

It can be particularly helpful to know 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 changes in thinking/behavior helps those who interact with the person. It can be difficult to know when to allow the person to act for him/herself and when to respond or act on his/her behalf. Knowing about thinking and behavior changes can make it easier for the caregiver to feel empowered to step in.

When insight and self-awareness are intact, advance directives to document wishes for care should be put in place in case insight and self-awareness diminish as the disease progresses. This is particularly important because cognitive and

behavioral impairments, like other symptoms of ALS, can advance with disease progression and worsen over time.

If insight and self-awareness are impaired, 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 a person with thinking/behavior impairment starts to withdraw or starts to become agitated in the face of requests for a 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 decision making towards the caregivers and family as the affected person will not and cannot be expected to change on his/her own behavior or show insight into offered interventions.

It is hard to know what he is thinking most days as he is a private person. I know things bother him when he says, 'No more ALS talk tonight.' Sometimes we spend too much time talking about what could happen instead of 'let's do this and to heck with ALS.'

Ellen, caregiver of person with ALS
(Contributed by The ALS Association North Carolina Chapter)

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.

Thinking and Behavior Symptoms and Their Impact on 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.

THE "PSEUDOBULBAR AFFECT"

Some people with ALS develop a symptom called "**pseudobulbar affect.**" They may cry or laugh at inappropriate times or notice how once they start feeling an emotion, it is difficult to suppress it and not express it. Sometimes there is 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. People with ALS can have pseudobulbar affect and no other cognitive, behavioral, or psychological symptoms. It can be easier on caregivers and family when the person experiences increased laughter rather than increased crying because we immediately associate crying with the feeling of sadness. However, when a person with pseudobulbar affect cries, it does not necessarily mean that a person is feeling anxious, sad, depressed, or emotionally distraught. **Pseudobulbar affect can be treated with medication if the symptoms become bothersome to the person with ALS or those around them. Please talk to your healthcare provider about the options that are available.**

Options for Addressing the Pseudobulbar Affect

- **Participate in testing of your thinking** (medical term: **cognitive screen or neuropsychiatric testing**). It will help you and your team to plan ahead.
- **Make a choice about who should speak on your behalf if you were to lose the ability to make your choices.** Find a trusted decision maker and let them know.
- **Make your wishes known.** The earlier you make your choices, the more confident you and your team will be about what you want.
- **Seek treatment for pseudobulbar affect if it bothers you.** You do not have to live with this symptom.

RECOMMENDATIONS FOR CAREGIVERS

- **Simplify communication with the affected person.** Break up sentences into short phrases. Ask yes/no questions. Slow down when speaking. If a person cannot speak, write down two choices and 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 and sustain it over time. 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 "underwater" 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 outside of dedicated ALS clinics may not have much experience with changes in thinking and behavior in 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.
- **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

It has been recognized that ALS can affect thinking or feeling. **Not everyone diagnosed with ALS will experience changes in thinking and behavior. It is assumed that about half the people with ALS will have impaired thinking and behavior, and the changes can range from mild to severe.** Knowing that this is potentially part of ALS 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 wishes are known and a trusted loved one can ensure care decisions are honored. Also, it is normal for caregivers to be challenged by changes in their loved one's thinking and behavior and additional support and resources 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 and what treatments may be effective when there are changes.





The following is a list of topics covered in the *Living with ALS* resource guides:

Resource Guide 1

What is ALS? An Introductory Resource Guide for Living with ALS

This resource guide provides an overview of ALS, what it is, and how it affects your body. It provides information on what kind of resources are available to help you deal with ALS more effectively.

Resource Guide 2

After the ALS Diagnosis: Coping with the “New Normal”

This resource guide addresses the psychological, emotional, and social issues that you must face when your life is affected by ALS. It provides information on how to cope with the many lifestyle changes and adjustments that occur when you live with ALS.

Resource Guide 3

Changes in Thinking and Behavior in ALS

This resource guide addresses how thinking and behavior may be affected by ALS and how these changes can impact disease course, symptom management, and decision making.

Resource Guide 4

Living with ALS: Planning and Making Decisions

This resource guide reviews areas where careful planning and decision making will be required and will provide you with resources to help you and your family plan for the future.

Resource Guide 5

Understanding Insurance and Benefits When You Have ALS

This resource guide provides strategies and helpful hints to better navigate health insurance and benefits. While understanding insurance and benefits may feel overwhelming, the guidelines outlined here should help simplify the process for you.

Resource Guide 6

Managing Symptoms of ALS

This resource guide discusses a variety of symptoms that may affect you when you have ALS. As the disease progresses, various functions may become affected and it is helpful to understand potential changes so that you know what to expect and how to manage these new changes and symptoms.

Resource Guide 7

Functioning When Mobility is Affected by ALS

This resource guide covers the range of mobility issues that occur with ALS. It discusses exercises to maximize your mobility, as well as how to adapt your home and activities of daily living to help you function more effectively.

Resource Guide 8

Adjusting to Swallowing Changes and Nutritional Management in ALS

This resource guide will help you understand how swallowing is affected by ALS and what you can do to maintain nutrition for energy and strength and to keep your airway open.

Resource Guide 9

Changes in Speech and Communication Solutions

This resource guide covers how speech can be affected by ALS and explores a variety of techniques, technologies, and devices available for improving communication. By maintaining communication with others, you continue to make a significant difference in their lives, while retaining control of your own.

Resource Guide 10

Adapting to Changes in Breathing When You Have ALS

This resource guide explains how breathing is affected by ALS. Specifically, it will teach you the basics of how the lungs function, the changes that will occur, and how to prepare for the decisions that will need to be made when the lungs need maximal assistance.

Resource Guide 11

Approaching End of Life in ALS

This resource guide examines thoughts and feelings about dying and end of life. Approaching end of life is difficult and support is critical to help sort out feelings, expectations, and plans. By talking to friends, family, professionals, and planning and communicating your wishes, you can help prepare for the best possible end-of-life phase.



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About The ALS Association

The ALS Association is the only national non-profit organization fighting Lou Gehrig's Disease on every front. By leading the way in global research, providing assistance for people with ALS through a nationwide network of chapters, coordinating multidisciplinary care through certified clinical care centers and fostering government partnerships, The Association builds hope and enhances quality of life while aggressively searching for new treatments and a cure.

For more information about The ALS Association, visit our website at www.alsa.org.