

COGNITION AND BEHAVIOR IN ALS

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ALS is primarily considered to be a disease that affects movement. However, we have recently learned that changes in cognition, or thinking, sometimes occur in the disease. Approximately one-third to one-half of patients with ALS experience changes in thinking. These are generally mild and do not cause concern and are noticed only with medical tests.

It appears that patients who present with speech or swallowing difficulty (bulbar-onset) develop cognitive difficulties earlier in the disease than patients whose weakness begins in the limbs. However, ALS patients whose symptoms begin in the limbs may also develop thinking difficulties.

ALS patients can also develop changes in behavior. Caregivers or loved ones notice changes in the way patients interact. Like cognitive changes, these are usually mild. However, in some cases, they can be more noticeable. As discussed below, some people seem to undergo pronounced personality changes and interact much differently than they once did.

COGNITIVE CHANGES SEEN IN ALS PATIENTS

“Cognition” refers to thinking, knowing, perceiving and understanding. It can be broken down into functions like attention, concentration, memory, language, organization of thoughts, planning and decision-making. We need to pay attention and concentrate in order to understand and learn. Attention and concentration are “gateway functions” to more complex abilities such as memory. Memory is very complex, involving imprinting information into the brain, filing it away, and pulling the information back out for later use. Many aspects of cognition are involved in simple daily tasks.

A number of different changes in thinking have been associated with ALS. Table 1 illustrates aspects of cognition and discusses which ones are affected in ALS.

Table 1: Cognitive Domains and Changes in ALS

Cognitive Domain	Purpose	Impaired in ALS?
Attention & Concentration	Focus on information coming into the brain	This can be impaired. A person may not listen to conversations as well or get distracted more easily.
Working Memory	Sustained focus while manipulating information	For example, this may involve mental calculations. This can be more difficult for patients with ALS.

Memory	Holding information in mind over short & long-term	Memory is usually not impaired in ALS. However, it may seem that someone's memory is poor because they have trouble paying attention, so information never gets to the memory center of the brain.
Language	Speech, expression of ideas	Language skills can be impaired. People may have trouble thinking of words, and spelling can be poor.
Flexibility of Thinking	Attending to more than one thing; weighing alternatives	This can be impaired in ALS. There may be trouble having a conversation while the TV is on, or there may be problems thinking of several possibilities at once. Also, they may have trouble benefiting from feedback from others.
Planning	Organization of intentions or ideas	Patients with ALS may have some problem with planning, which may be related to inflexible thinking.
Visual-Spatial Skills	Perceiving visual information	This is generally not impaired in people with ALS
Judgment & Insight	Making good decisions and understanding one's situation	This can be impaired in some people with ALS. Those with behavioral changes may be at greater risk for problems in this area.

BEHAVIORAL CHANGES ASSOCIATED WITH ALS

Unlike cognitive changes, which are often subtle, behavioral changes can be prominent. Like cognitive changes, it is often a family member who notices personality changes before the patient.

It is important to remember that these changes do not occur in all people with ALS and people without ALS can also have days when they are irritable or less polite than usual. Changes in behavior are only considered problematic when they occur on a consistent basis and disrupt work, relationships, home life or health.

Table 2: Behavioral Changes in ALS

Possible Behavioral Problems	Example
Apathy: A lack of initiative or interest in people and/or activities. There may be a lack of spontaneous behavior or an appearance of low motivation.	An apathetic person fails to engage in conversations or to become involved in things that interest them (like hobbies). They might watch television for hours on end. Apathy can be mistaken for depression, but if asked, the person with apathy generally will not report sadness.

Disinhibition: Loss of a usual and appropriate sense of social inhibition or a lowering of self-control.	A person who is disinhibited may lack social graces even though they were previously very socially aware and well controlled. They may say inappropriate things, or grab for something in a rude manner.
Lack of Empathy: A decreased awareness of the feelings or experiences of others	A person with diminished empathy may show a lack of emotional response to others or a blank facial expression, even when someone is very upset. They appear to “not get it” emotionally.
Self-Centeredness: Focusing on one’s own needs without consideration of others	Someone who is self-centered continually talks about their problems or concerns and does not consider the needs of others. This is related to decreased empathy.
Decreased Insight: Reduced awareness of one’s own situation.	An individual with decreased insight does not understand how they have changed or that their behaviors are inappropriate.

MORE ADVANCED DEFICITS-FRONTOTEMPORAL DEMENTIA

In a percentage of patients with ALS, impairment is more extensive and fits a profile known as “Frontotemporal Dementia (FTD)”. FTD reflects a combination of cognitive and behavioral/personality changes. A patient with FTD may have significant problems with language skills leaving them unable to communicate effectively with others. The behavioral problems associated with FTD can become quite extreme. Patients with FTD may have little control over their impulses, becoming aggressive or explosive. Conversely, they may become severely withdrawn and apathetic. FTD causes patients to have significant difficulty functioning in normal social situations or living independently. This is a condition that is progressively disabling, but if it is identified early, treatments can be provided. Accommodations such as techniques to manage difficult behaviors can also be made so that problems are minimized.

The term dementia is often associated with Alzheimer’s disease (AD). However, the changes in ALS and FTD are *not* the same as AD. Alzheimer’s disease affects many abilities but it primarily disturbs memory. Individuals with AD have great difficulty learning and retaining new information, and eventually forget things like the names of their loved ones or where they live. The changes associated with ALS generally do not affect memory. People with ALS may have slight problems with memory if their concentration is worse, but once the information gets in, patients with ALS generally do not forget it. Poor concentration can be a function of depression or a specific change in cognition.

TREATMENT OF COGNITIVE AND BEHAVIORAL PROBLEMS IN ALS

One approach to treating cognitive and behavioral changes involves medication. Some medications help people feel more alert, which helps with depression or apathy. There are also

medications that are calming, which can help people who are anxious, socially inappropriate or irritable. Medications targeted specifically for Alzheimer’s Disease, have been used in other diseases but have not been widely studied in ALS.

Another way to manage cognitive and behavioral changes is to modify the environment. For people with cognitive changes, it is helpful if their home is structured and organized. For example, establishing specific locations for frequently used things such as keys, wallet or glasses is helpful. Occupational therapists are very helpful in providing resources and strategies for adapting the home. Another means of adding structure includes organizing daily information by using a calendar or notes.

Communication is another important issue. Again, minimizing distractions is recommended. Distractions can cause individuals to lose their focus on a topic and can also cause agitation or irritability. Giving too much information when talking to patients with cognitive problems, leads to information overload and decreased concentration.

OTHER FACTORS AFFECTING BEHAVIOR: DEPRESSION AND EMOTIONAL LABILITY

Depression

Depression is a sense of feeling sad, down or tearful. These feelings can be quite normal if they occur intermittently, but if they last for two weeks or more and do not seem to lessen, then depression may be present. Approximately 5-7% of all people suffer from depression at some time in their lives, with women being at higher risk. In ALS, about 20 - 30% of patients report persistent depression, and many tend to feel most depressed after diagnosis.

<p>Clinical Depression is NOT:</p> <ul style="list-style-type: none"> • Occasional tearfulness • Being in a bad mood • Sadness following upsetting news • Not communicating much with others • Emotional Lability: Excessive crying/laughter at inappropriate times. 	<p>Clinical Depression IS:</p> <p>At least <u>2 weeks</u> of feeling down, sad or blue on most days, along with several of the following:</p> <ul style="list-style-type: none"> • Losing interest or pleasure in things that usually bring pleasure • Notable changes in appetite • Losing or gaining weight not associated with feeding tube or other ALS-related conditions • Sleep changes <ul style="list-style-type: none"> - waking in the middle of the night without being able to fall back asleep, or - being unable to fall asleep • Excessive fatigue and restlessness
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	<ul style="list-style-type: none"> • Thoughts of suicide • Severe enough to impair your ability to work, take care of daily tasks or get along with your friends and family.
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Depression can also indirectly affect thinking. It can become harder to concentrate, make decisions, or initiate activities. These problems are similar to the problems associated with ALS. Clarification from health care professionals is important so you know whether depression or thinking changes are causing difficulty with concentration.

Depression is treated with antidepressant medications and psychotherapy (talk therapy). These are effective when offered together. Psychiatrists, primary care physicians and neurologists are three of the most common physicians that can prescribe medications to treat depression. Psychologists, psychiatrists, social workers, and other counselors are trained to treat depression using psychotherapy.

Alcohol is not an effective means for trying to cope with depression. Abusing alcohol may temporarily decrease the emotional pain of depression but it actually makes the depression worse and impairs thinking and coping. Others try to handle depression by withdrawing from others, but this is also a poor way to manage emotional problems.

Emotional Lability

Emotional lability is also known as “pseudobulbar affect”, “emotional incontinence” or “pathological laughing and crying”. It is a neurological symptom that occurs in ALS, usually in people who have speech and swallowing difficulty. It is characterized by sudden outbursts of uncontrollable laughing or crying. These episodes are not provoked by humorous or sad conditions, but often occur spontaneously and seem quite out of context. These episodes are difficult to control and are not symptoms of depression or an other psychiatric condition. Several medications, especially antidepressants, can be used effectively to minimize this symptom.