ALS and Cognitive Changes: A Guide for Patients and Families

Overview

A link between ALS and cognitive dysfunction (problems with thinking and/or behavior) was first noted in the late 1800's. However, it's only been in the last few years that new research has shed important light on how these two conditions might be related. What was once considered a rare link between two different diseases has now been identified as a more common combination. Knowledge about the relationship between ALS and cognitive changes is growing and changing. More research is planned to increase medical understanding and to help in the management of patient care.

There is a very wide range of what patients with ALS may experience – from no cognitive changes at all, to mild or moderate difficulties, and in some cases, more severe problems.

- Many people with ALS will have no changes in behavior or thinking.
- Recent studies suggest that it is not uncommon for patients with ALS to have mild-to-moderate thinking or behavioral problems. In some cases, the thought or behavior problems may be so mild and subtle that they aren't noticed by friends or family members and don't interrupt daily activities, and are only recognized through neurologic testing. For other, the symptoms (described below) are more noticeable and do impact daily activities.
- A sizable minority of people with ALS may have a more serious form of these problems. It's called frontotemporal lobar dementia (FTLD). The location of the degeneration of neurons that causes the problems happens in the front and side parts of the brain.

Because for a long time it was believed that ALS did **not** affect a person's mental processes, health care professionals are now taking another look at some commonly-held approaches to managing the care of people with ALS. For patients and families, just knowing that problems with cognitive function and behavior may be a part of the ALS disease process can be helpful to explain what they may already be observing. There are a wide range of approaches families can take to better manage problems related to these problems. It's important to know that the cognitive changes that can be experienced with ALS are due to degeneration of brain cells, and are not a psychiatric illness.

What Are Cognitive Impairment (CI) and FTLD?

Cognitive Impairment (CI) and Frontotemporal Lobar Dementia (or FTLD) are a mild to severe continuum of a brain disease that causes particular brain cells to slowly, consistently die. This illness causes the patient to have personality changes, language difficulty and/or behavioral disturbances. Some people with ALS show only limited signs of cognitive or behavioral changes (CI), while others develop symptoms that may lead to a diagnosis of FTLD. The neuron degeneration and changes of CI and FTLD have been identified using Magnetic Resonance Imaging (MRI) technology.

The behavior and thinking problems experienced in the more severe FTLD are different from the dementia of Alzheimer's disease and presents different signs and symptoms. For example, people with FTLD typically develop behavior symptoms first, while people with Alzheimer's disease usually develop memory problems first. Many of those with FTLD may not experience memory problems at all.

Symptoms of CI and FTLD

Sometimes the symptoms of ALS can be confused with symptoms of CI or FTLD. Medications, depression, or problems with respiratory function can all cause some of the same symptoms that CI or FTLD can cause. It's important for a patient's symptoms to be evaluated by an ALS health care provider to find out the cause of the problems. Because people with ALS typically experience a steady decrease in their ability to speak, swallow, move and perform activities of daily living, it can be easy to miss noticing some common thinking problems.

The main symptoms of CI and FTLD have to do with behavior and personality changes and they develop gradually over time. Some changes in personality might include the following:

- Less sensitive to the need of others
- Behave in ways that are embarrassing, inappropriate, and uncharacteristic
- "Too cheerful" and may seem childish
- More withdrawn and need prompting to do most things
- Changes in behavior such as eating lots of sweets and stuffing food in their mouth without swallowing
- Fixated on one idea or activity
- More aggressive

Other symptoms of CI or FTLD have more to do with thinking. For example, many people with cognitive impairment may not recognize that they have any problems or changes, and as a result make some bad decisions about how to take care of themselves, perform their work, or manage their health. They may have difficulty solving problems, especially if the problem is something new such as using a new piece of equipment, or if it involves more than a few steps.
Some people lose insight into their illness, and so would not be able to discuss the cognitive and emotional changes they are experiencing.

Patients may not care much about or recognize their problems, they may not be able to focus attention, or they may become overly agitated. "Executive function" is a thinking skill that is difficult for ALS patients with cognitive problems. "Executive function" refers to a type of complex thinking when people handle many types of information at the same time; such when one drives a car through busy streets at rush hour. This skill is also used when patients are asked to make complex decisions about life-style changes and medical interventions when the patients is given detailed information about changes in speaking, swallowing, breathing, and muscle control.

Each patient with these cognitive changes will have different kinds of problems.

- Some with only changes in behavior and personality,
- Some with only thinking problems, and
- Others with both types of problems.

Because of their thinking problems, some patients are not able to understand the changes that are happening to them. Family members too may see the cognitive changes as just frustration or anger with the illness itself or with the caregiver.

Risk Factors for CI and FTLD

Patients over the age of 60, who had bulbar onset ALS, poor breathing, or a family history of dementia, may be more likely to have cognitive impairment of FTLD.

Diagnosis of CI and FTLD

A diagnosis of CI or FTLD is made when particular behaviors are seen in patients. People with FTD may have problems interacting with others socially, have less emotional reactions to things that would normally upset people, and they often do not understand what is happening to them.

To diagnose the presence of this brain disease, your physician will refer you to a neuropsychologist for a neuropsychological and neurobehavioral evaluation. This evaluation will take several hours and are not blood tests or like other physical diagnostic tests. In the ALS clinic, a few tests may be given to patients to see if they should have this type of evaluation. Patients may be asked to say a list of words as quickly as they can and caregivers may be asked a series of questions about the patient's behavior.

Learning that the Patient Has Cognitive Changes

If doctors suggest that CI or FTLD is present, the diagnosis is sometimes a relief to the family, because is often explains problems with thinking or behavior that were previously misunderstood. Some family members say that learning more about CI and FTLD helps explain behaviors such as aggressive behavior or a patient's lack of concern about what is happening to them or their families. Naturally, this can be a very difficult time for patients and family members.

Things that can help patients and families cope with the thinking and behavior changes the patient may experience, and may be able to make the situation a bit easier:

- Become informed about what is happening.

- Talk frankly with the patient's health care providers about the symptoms, possible treatments and ways to help the patient. Ask about things caregivers and family members can do to help the specific thinking or behavior problems their loved one is having.

- Get support or counseling.

- Know that developing cognitive changes have nothing to do with anything the patient or family may have done, or not done.

Length of Survival and Acceptance of Medical Treatments

Studies suggest that, for reasons that are not yet completely understood, patients with both ALS and CI or FTLD may have a shorter lifespan than those patients with ALS alone. Some health care providers believe that this problem is related to their observations that ALS patients with cognitive and behavioral problems do not follow their doctor's recommendations as often as patients without CI or FTLD.

This shortened life span may affect families, especially as they involve patients in treatment decisions and in legal matters, such as advance directives, wills, and related documents. These challenges may occur sooner than family members had anticipated and the loved one with ALS may not want to participate in or accept the important decisions that need to be made.

Recommendations for Care and Support

Ways you may be able to help those with cognitive or behavioral problems:

- Remember that the patient has these problems because their brain is changing. The patient can not simply "fix" the problems by trying harder. Equally important, it is not the caregiver's fault. The caregiver and family can't "fix" the problem by trying to work faster or harder. Caregivers and families are not to blame if patient's behavior or thought problems worsen or interfere with socialization and daily activities.

- Use simple and straightforward language and communicate clearly and directly.

- Supervise the patient's eating more closely. Those with poor swallowing may have trouble following medical advice to limit solid foods, or they may place too much food in their mouth at one time. Some patients may eat more food than they need.

- Help your loved one make decisions about their care. The patient may not understand why they need to use walkers, wheelchairs, breathing equipment or feeding tubes.

- Supervise walking. Patients may make decisions too quickly, without remembering to be careful. They may walk in dangerous places, walk too far, or forget to use equipment such as a walker.

- Try to build an atmosphere of comfort and love, with a calm, structured and orderly environment.

Medications for FTLD with ALS:

While physicians cannot cure these cognitive and behavioral problems today, some medicines may provide a degree of relief from symptoms. Because each patient has their own, unique type of problems, your doctor will choose which medicines will be helpful for each individual patient.

Summary

For years, patients and families have taken some level of comfort from the thought that although ALS poses unique and serious physical challenges, the one issue they were not likely to have to face was a decrease in cognitive function. Unfortunately, research now suggests that this may not be true for all ALS patients.

The diagnosis and treatment for CI and FTLD with ALS is improving. Researchers are hopeful that the discovery of the connection between ALS and CI and FTLD among some patients may speed the understanding of the cause of ALS and provide important clues to the puzzle ALS still presents.

In applying any of this information to the care of a particular individual, we recommend talking with their healthcare professional first.

Resources

The ALS Association
Patient Services Department
27001 Agoura Road, Suite 250
Calabasas Hills, CA 91301
Telephone: 818-880-9007 / Fax: 818-880-9006
Toll-free Information and Referral Service: 800-782-4747, or 818-880-9007
Email: alsainfo@national.org
www.alsa.org
Contact your local ALS Association Chapter for programs, services and resources to help patients and families with the day-to-day challenges of living with ALS and cognitive impairment. Locate the chapter closest to you through your local telephone directory, The ALS Association's web site, or by calling the toll-free number listed above.

Family Caregiver Alliance
690 Market Street, Suite 600
San Francisco, CA 94104
Telephone: 415-434-3388 / 800-445-8106
www.caregiver.org
Email: info@caregiver.org
The Family Caregiver Alliance offers a range of resources, including a "Fact Sheet on Behavior Management Strategies" for dementia available on their web site.

fyi...
for your information

The ALS Association thanks J. Murphy and M. Lyon for their contributions to this FYI.

Created 2005
Updated 2008