

CARE OF ALS: DRUG, SYMPTOM, NUTRITIONAL, AND BREATHING THERAPIES



This fact sheet may help you understand which treatment choices are best for managing symptoms and problems with nutrition and breathing in people with amyotrophic lateral sclerosis (ALS), sometimes referred to as PALS. For information on multidisciplinary care and management of behavioral and thinking problems in people with ALS, please read the AAN Summary of Evidence-based Guideline for Patients and their Families on *Care of ALS: Multidisciplinary Care and Management of Behavioral and Thinking Problems*.

Neurologists from the American Academy of Neurology are doctors who identify and treat diseases of the brain and nervous system. The following evidence-based information* is provided by experts who carefully reviewed all available scientific studies on the care of people with ALS.

What is ALS?

ALS is a disease that affects your ability to control voluntary muscles. It is also known as Lou Gehrig's disease. In ALS, certain nerve cells in the brain and spinal cord become damaged. The affected cells are called motor neurons. These nerve cells send signals to the muscles that you can control, such as the muscles in your arms and legs. The nerves that control automatic actions in the body, such as your heartbeat and digestion, are not affected.

People with ALS lose muscle strength and control. Eventually they are not able to stand or walk, or use their hands and arms; they also have difficulty breathing and swallowing.

The cause of ALS is not yet known, and researchers continue to look for one.

Is there a cure for ALS? Will I die from it?

ALS is a serious disease. However, it is important to know that several therapies are now available to make daily life easier and more comfortable.

ALS progresses quickly and eventually leads to death. Most people with ALS die within three to five years from the onset of their first symptoms.

However, if you are diagnosed with ALS, you should know that it is possible to live longer and with some relief from symptoms. About 10 percent of people with ALS survive for 10 years or longer. Also, the available treatments can make daily life easier and more comfortable.

My doctor says I have ALS. Are there any medications I can take to help me live longer or to treat my symptoms?

As of September 2009, riluzole is the only medication approved by the U.S. Food and Drug Administration (FDA) to treat ALS. Riluzole has a modest effect on slowing the rate at which the disease worsens. Strong evidence shows that riluzole can prolong survival by two to three months. There is also strong evidence that this medication is most helpful when started early in the course of the disease. Over the past 10 years, people who have used riluzole have reported few side effects. The most common side effects are fatigue and nausea (upset stomach). It is important to know that riluzole is an expensive medication. People who are newly diagnosed with ALS should discuss with their neurologists whether beginning riluzole treatment would be helpful for them.

There are medications available that can be very helpful in treating some symptoms of ALS. One symptom, uncontrolled laughing or crying, can be embarrassing and hard to manage. A person with ALS at times may laugh longer than expected at something that is only mildly funny. He or she also may laugh at something typically considered sad or cry in response to something funny. There is good evidence that the medications dextromethorphan (DM) and quinidine (Q) will lessen episodes of uncontrolled laughing or crying—both how severe they are and how often they happen. The side effects associated with DM and Q are dizziness, nausea, and sleepiness, so some people stop taking them. These medications are currently awaiting FDA approval.

Another symptom, drooling, happens because of swallowing problems. Drooling can be embarrassing, and trouble with swallowing the drool can be a health risk. The excess saliva can be accidentally breathed in, which in some cases contributes to pneumonia.

There is good evidence that injections of the medication botulinum toxin type B (BTxB) can help with drooling. The medication is injected into the glands near the jaw that make saliva. No serious side effects have been reported from use of BTxB.

Are there other ways of treating these symptoms?

You can get radiation therapy for drooling if BTxB does not give relief. There is weak evidence that small amounts of radiation therapy might help. The side effects of this therapy are sore throat, nausea, and reddening of the skin. Ask your doctor whether radiation therapy would be a good choice for you.

Also, services such as physical therapy, occupational therapy, and speech and language therapy can help you live better and more fully with your symptoms. For more information, read the AAN fact sheet *Care of ALS: Multidisciplinary Care and Management of Behavioral and Thinking Problems*.

Will having trouble swallowing affect my health? How can this problem be treated?

When you have problems swallowing, eating becomes very difficult. It is common to begin to choke and accidentally breathe in food you are trying to eat. Eventually weight loss and dehydration (not enough fluid in your system) occur. It is important to manage your swallowing problem so that you maintain good nutrition. Changing the texture of your food, taking smaller bites, and tucking your chin while you swallow can help.

You can also use a feeding tube called a percutaneous endoscopic gastrostomy (PEG) tube or radiologically inserted gastrostomy (RIG) tube. These types of tubes are placed in the stomach. There is good evidence that a PEG or RIG tube will likely help stabilize body weight and prolong survival.

How can problems with breathing be treated?

Breathing problems develop in people with ALS because of weakening of the muscles that help with breathing. Treating breathing problems is very important. The most common cause of death from ALS is not getting enough oxygen due to breathing problems. It can be hard to know if you have a breathing problem because you may not notice it until it becomes serious. Talk to your doctor about available tests to detect a breathing problem.

If your doctor discovers that you have a breathing problem, he or she may recommend that you use an assisted-breathing device, also known as a noninvasive ventilation (NIV) device. Good evidence shows that using an NIV device will likely lengthen survival and slow down development of breathing problems. There is weak evidence that shows use of an NIV device will improve quality of life.

Muscle weakness that affects breathing can also affect the ability to cough. If your cough is weak, you might have trouble clearing mucus and other fluids from your throat, which may lead to a chest infection. Assisted-coughing devices, also known as mechanical insufflation/exsufflation (MIE) devices, are available to help with coughing. Weak evidence shows that using an MIE device may help clear fluids from the throat, especially during a chest infection.

This statement is provided as an educational service of the American Academy of Neurology. It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The AAN recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved.

*After the experts review all of the published research studies, they describe the strength of the evidence supporting each recommendation:

Strong evidence = more than one high-quality scientific study

Good evidence = at least one high-quality scientific study or two or more studies of a lesser quality

Weak evidence = the studies, while supportive, are weak in design or strength of the findings

Not enough evidence = either different studies have come to conflicting results or there are no studies of reasonable quality

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