

Treating the Symptoms of ALS

People with ALS frequently complain that, after giving them the diagnosis, their doctors do not want to see them, do not want to talk to them, and do not know how to comfort them. In fact, many physicians may feel powerless in managing a person with ALS. The important point is that, although ALS cannot be cured, it *can* be effectively treated. In this chapter, we will discuss both medications to slow the progression of the disease and also the treatment of specific symptoms. If you receive your care at an ALS clinic, these treatments will be familiar to your physician and he will inform you of ways in which he can help you cope with your disease. If you receive your ALS care from a community physician, this chapter might be useful to share with your treating physician, as it may provide him with ideas as to how better to help you manage your illness. You have the right to expect your doctor to take the time to become informed about your specific problems with ALS. At each appointment, you have the opportunity to teach your physician more about ALS and how it affects you. Your physician has the responsibility to help prepare you for the future so that the impact of the disease on your life will be lessened.

Treatment to Slow the Progression of ALS

People with ALS are primarily interested in pursuing treatment once the diagnosis of ALS has been established. The only specific treatment for ALS that is approved by the U.S. Food and Drug Administration (FDA) at this time is riluzole (Rilutek®). Two large placebo-controlled double-blind trials demonstrated that riluzole modestly extends survival for people with ALS (see Chapter 3). Further analysis of the data suggested that patients

treated with riluzole stayed in milder ALS disease states longer than patients who were not treated.

Treating Specific Symptoms

Table 6-1 summarizes the medications commonly used to treat the symptoms of ALS.

Weakness

Although the progressive weakness of ALS cannot be stopped or reversed, there are adaptive strategies. Occupational and physical therapists can provide numerous assistive devices to help with feeding, dressing, walking, and other activities of daily living (see Chapters 4 and 9).

Muscle Aches, Cramps, and Spasms

A muscle cramp is a sudden, unintended muscle contraction that may occur in the arms, legs, chest, back, abdomen, jaw, or throat. Cramps are caused by overstimulation of the nerves of a weakened muscle. These contractions may be extremely painful and prolonged. You may experience knotting in the muscle and abnormal posture until the cramp passes. You can help manage cramps by maintaining a proper diet, drinking plenty of water, and avoiding overexertion of your weaker muscles. Stretching is also a very effective way of stopping a muscle cramp. Medication can be prescribed when cramps are particularly frequent and severe.

Spasticity

Spasticity is a change in the *resting tone* of your muscles that leaves them feeling tight and stiff. The legs become difficult to bend, and walking becomes unbalanced and slow when spasticity is severe. The legs will not relax when sitting and will have a tendency to straighten at the knee. This stiff feeling can make it difficult to fall asleep at night and remain comfortable throughout the night. Many medications are available to relax this

TABLE 6-1

Commonly Used Medications for Symptomatic ALS Treatment

Symptom	Medication
Muscle cramps and spasms	Baclofen, tizanidine (Zanaflex [®]), quinine sulfate (quinine)
Excessive laughter or crying	Tricyclic antidepressants (Amitriptyline [®]), selective serotonin reuptake inhibitors (Lexapro [®] , Celexa [®] , Zoloft [®] , Prozac [®]) valproate (Depakote [®]), lithium (Lithobid [®])
Spasticity (stiffness of limbs)	Baclofen, tizanidine (Zanaflex [®]), benzodiazepines (Valium [®])
Urinary urgency or frequency	Oxybutynin (Ditropan [®])
Excessive saliva	Tricyclic antidepressants (Amitriptyline [®]), glycopyrrolate (Robinul [®]), scopolamine (TransDerm [®]), botulinum toxin injection (Botox [®])
Thick phlegm/postnasal drip	Guaifenesin (Robitussin [®] , Humibid [®]), nebulizer treatments
Laryngospasm (throat-closing spasm)	Benzodiazepines (Valium [®] , Klonopin [®] , Ativan [®])
Jaw quivering/clenching of the teeth	Benzodiazepines (Valium [®] , Klonopin [®] , Ativan [®])
Nasal congestion	Steroid nasal spray (Beconase [®] , Nasonex [®])
Acid reflux	Omeprazole (Prilosec [®]), famotidine (Pepcid [®]), ranitidine (Zantac [®]), cimetidine (Tagamet [®]), metoclopramide (Reglan [®])
Insomnia	Tricyclic antidepressants (Trazodone [®]), zolpidem (Ambien [®]), temazepam (Restoril [®])
Depression/anxiety	Selective serotonin reuptake inhibitors (Prozac [®] , Paxil [®] , Zoloft [®] , Celexa [®] , Lexapro [®]), venlafaxine (Effexor [®]), bupropion (Wellbutrin [®]), mirtazapine (Remeron [®])
Shortness of breath	Morphine elixir
Pain	Nonsteroidal anti-inflammatory (Motrin [®] , Celebrex [®] , Vioxx [®]), pain medication (Tylenol [®] , Darvon [®] , aspirin), narcotics (Vicodin [®] , morphine, Oxycontin [®])
Nausea	THC (Marinol [®]), prochlorperazine (Compazine [®])
Constipation	Stool softeners (Colace [®]), laxatives (Senekot [®] , Ducolax [®]), fiber (Metamucil [®]), enemas
Agitation/anxiety	Benzodiazepines (Ativan [®] , Xanax [®] , Valium [®])
Terminal agitation	Morphine, atypical neuroleptics (Seroquel [®] , Zyprexa [®]), thiorazine (Thioridazine [®]) (if sedation desired)

increased muscle tone or stiffness and restore muscles to a more natural state. Care must be taken not to use high doses of these medications, because excessive muscle relaxation or weakness could result.

Fatigue

Generalized fatigue and exhaustion are common features of ALS. As nerve cells die, the remaining ones send signals to activate the otherwise unused muscle and a single surviving nerve cell may be doing a hundred times its normal workload. This may result in temporary exhaustion of overworked nerve cells. Thus, there may be times when you can perform a task, such as climbing stairs, only when you have rested beforehand. You should pace yourself wisely in terms of energy expenditure throughout the day and the week. Various medications have been tried for fatigue with little success and are therefore not recommended. When persistent overwhelming fatigue is a problem, your quality of sleep should be evaluated, because ineffective sleep and also depression are common causes of daytime fatigue.

Sleep Disturbance

Sleep disturbance can erode quality of life and cause other difficulties, such as depression and fatigue. Many factors can contribute to an inability to sleep. Sleep disorders in ALS may result from depression and anxiety, as well as nighttime breathing disturbances due to sleep apnea or weakened breathing muscles (see Chapter 10). If you are not sleeping well, you and your doctor should try to determine why. The cause could vary from simple insomnia to anxiety and depression or breathing problems. The best treatment for problems with sleep will be the one that gets at the specific problem that prevents you from sleeping.

Excessive Laughter or Crying

People with ALS sometimes develop difficulty controlling their emotions and will cry or laugh inappropriately or excessively. This is thought to be

caused by the loss of motor control over the brain centers involved in laughing and crying. These symptoms are embarrassing and they are often not recognized as being part of ALS. Antidepressants and the other medications listed in Table 6-1 are effective in treating these unwanted displays of emotion.

Urinary Urgency or Frequency

Although ALS usually does not involve the bladder muscles in the early phases, people with ALS frequently develop an irritable bladder with urinary urgency and frequency. This may be due to the loss of motor control over the brain centers for urination. In addition, urinary tract infections will also increase bladder spasms and frequency of urination. Therefore, when there is a marked increase in urinary frequency, an analysis of a urine sample should be performed, and any infection should be treated with antibiotics. In men, the prostate gland may be enlarged, resulting in the need to urinate frequently. Medication may be helpful to relax the bladder and lessen urinary symptoms if no infection is present.

Swelling of Hands and Feet

Swelling frequently occurs in a very weak limb due to failure of the muscle pumping action that increases blood return to the heart. Passive range of motion, elevation of paralyzed limbs, and compression hose are helpful. When swelling in one leg is painful or fails to decrease after overnight elevation, a blood clot or deep vein thrombosis (DVT) should be suspected and evaluated immediately because of the risk of the clot traveling to the lungs (pulmonary embolism). You should discuss blood clot prevention with your physician prior to prolonged periods of inactivity, such as airplane travel.

Excessive Salivation

Excessive saliva, a common feature of ALS, results in increased drooling, choking, or coughing. The problem is not due to overproduction of saliva,

but to decreased swallowing. Saliva is not swallowed automatically by those with ALS, and you must consciously swallow saliva to compensate for this. Excess saliva may be further increased by anxiety, hunger, and acid reflux. There are many medications to decrease saliva (see Table 6-1, page 93), and you can request them from your physician. Surgery and radiotherapy have both been tried for excess secretions but often result in excessively thickened secretions. Botulinum toxin (Botox®) injections into the salivary glands have very effectively reduced excess saliva in many people with ALS when medications are no longer effective. This treatment lasts for approximately 3 months and may be repeated when needed.

Thick Phlegm/Postnasal Drip

People with ALS may develop the habit of breathing through the mouth because of fatigue of the jaw muscles or nasal congestion. Breathing through the mouth causes the saliva to dry out and thicken. Furthermore, medications taken to reduce drooling can cause excessive dryness and thick secretions, which can result in postnasal drip, chronic cough, or a need to clear the throat. Certain medications have been reported to thin secretions when added to antisaliva regimens. Nonprescription cough syrups may be helpful in thinning saliva. Chicken broth and hot tea, as well as the use of room humidifiers, oral suction machines, and aerosolized breathing treatments can also help. When allergies contribute to postnasal drip, antihistamines and steroid nasal inhalers may also be useful.

Jaw Quivering and Clenching of the Teeth

Some people with ALS may experience an uncomfortable tightening or chattering of the jaw due to cold, anxiety, or pain. Medications may be helpful in relieving these symptoms as well.

Laryngospasm (Tightening of the Throat)

Laryngospasm is an abrupt and prolonged closure of the vocal cords resulting in sudden gasping for breath and wheezing. This can cause panic

because of the fear of suffocation. Laryngospasm may also occur with increased emotion or with exposure to smoke, strong smells, alcohol, cold or rapid bursts of air, and even spicy foods. It can also occur with sinus or postnasal drip, as well as acid reflux. Laryngospasm normally clears spontaneously in a few seconds, but can be more immediately relieved by breathing through the nose and repetitively swallowing. Possible triggers of laryngospasm should be eliminated and a trial of antacids instituted. Understanding the nature of this process can usually help to calm the panic that aggravates laryngospasm.

Acid Reflux

Acid reflux, also known as gastroesophageal reflux disease (GERD), is a common condition in people with ALS. It is due to weakness of the diaphragm muscles involved in breathing, which normally form a tight band around the opening to the stomach to keep the acids down. The signs and symptoms may include heartburn, acid taste, throat irritation, chest pain, hoarseness, shortness of breath, nausea, insomnia, and even spasms of the larynx. These symptoms are thought to be caused by the reflux of stomach acid into the lower esophagus. Caffeine, spicy foods, overeating, and diaphragm weakness all increase acid reflux into the esophagus. Particular care in managing this problem is necessary if a feeding tube is used because the stomach may be easily overfilled. Medications to increase clearance of foods from the stomach (metoclopramide) as well as antacids (ranitidine hydrochloride, cimetidine, famotidine, and omeprazole) are quite effective.

Nasal Congestion

The muscles in the nose and mouth may be weakened, resulting in a failure to elevate and open the nostrils, the upper airways, and the Eustacian tubes, which connect to the ears. The nasal airways can be opened effectively by applying nasal tape across the bridge of the nose to open nasal passages, as used by athletes. Steroid nasal sprays take several days to take effect but can be very helpful in relieving congestion.

Constipation

Constipation is common in ALS and, if untreated, can result in hours spent on the toilet with abdominal pain and bloating. Constipation in ALS is caused by decreased fluid, inadequate diet, lack of exercise, and a reduced ability to bear down with the abdominal muscles. Proper management is essential because hospitalization for bowel obstruction may be necessary if this condition persists. Medications taken to control excessive saliva and pain can contribute to constipation and should be decreased if possible or discontinued when not necessary. An excellent dietary recipe to normalize bowel movement is “power pudding,” which consists of equal parts of prunes, prune juice, applesauce, and bran. Two tablespoons with each meal and at bedtime, along with adequate fluid intake and fruits and vegetables in the diet, helps in maintaining normal bowel movements. Stool softeners, laxatives, and periodic enemas should be used liberally, when necessary.

Depression and Anxiety

Depression is common in anyone with increasing physical disability and should be aggressively treated. Depression is underdiagnosed and undertreated in ALS, and it has a negative effect on the quality of life of both patients and their families. Newer antidepressants (the selective serotonin reuptake inhibitors) are preferable to the older types of antidepressants (such as amitriptyline) because of their greater efficacy and lesser side effects. When effective, antidepressants should be continued for at least 6 months to one year and then slowly tapered as indicated. Buspirone and benzodiazepines can be used on an as-needed basis when anxiety and agitation are the main problems. Buspirone is preferable because it does not suppress the muscles involved in breathing.

Difficulty in Swallowing

Weakness and incoordination of the mouth and throat muscles can result in swallowing problems. A swallowing evaluation by a speech therapist can be helpful in determining which foods cause the greatest difficulty (see

Chapter 7). For mild swallowing problems, you should tuck your chin down, swallow two to three times per mouthful of food, avoid foods that cause the greatest difficulty, and perform a clearing cough after each swallow. When swallowing difficulty results in weight loss or when a patient becomes fatigued in attempts to consume a meal, a percutaneous gastrostomy (PEG) should be considered. The procedure should be carried out before excess weight loss occurs and while the patient has good breathing function, because the risk of complications increases in weaker patients. They should be fed only in an upright position and should avoid bending or lying flat for at least 1 hour after eating, to prevent food from entering the lungs. In many instances, medicines such as metoclopramide may be added to aid emptying of the stomach. Most people with a PEG prefer feedings at regular mealtimes to leave time available for other activities. A variety of feeding formulas are available. In general, concentrated formulas with high fat and sugar content are harder to digest and may result in cramping and diarrhea. Less concentrated solutions are preferable and may be specifically selected to meet individual needs (i.e., high-fiber formulas for constipation; high protein, high lipid, low carbohydrate formulas for patients with shortness of breath) (see Chapter 7).

Slurred Speech

Slurred, slow, or strangulated speech is caused by incoordination and weakness of the lips, tongue, and throat muscles. People with speech problems should be evaluated by a speech therapist and alternative communication methods explored (see Chapter 8). Some form of communication can usually be established and must be sought. It is essential that the family members recognize and respect the patient's need to communicate, even when it is time-consuming and frustrating.

Shortness of Breath

Breathing problems may arise from many different causes in ALS. For this reason, breathing difficulties should be evaluated by a physician familiar with both breathing problems and ALS (see Chapter 10).

Pain and Pressure Sores

Although pain is not generally thought to be a feature of ALS, it is extremely common late in the course of disease. Pain may be due to muscle cramping or joint changes such as hip dislocation. Pain can also occur when there is prolonged immobility of paralyzed limbs, such as when a caregiver is not available to reposition the person frequently. A pressure sore may develop if the body has been in the same position for many hours. Pressure sores are particularly apt to develop over bony areas, such as the tip of the spine. Alternating airflow mattresses and air and gel cushions can reduce pressure over these tender spots. Mild pain associated with pressure sores may be relieved by ibuprofen or acetaminophen, but narcotic medications may be needed for more severe pain.

Alternative Treatment

People with ALS are often dissatisfied (as are their physicians) with the specific treatment offered by conventional medicine to slow the disease process and many patients seek alternative solutions to their disease. These holistic approaches may benefit the patient greatly in affording a sense of mastery over the disease. Nutritional regimens that stress the role of antioxidant vitamins in postponing symptom manifestation have some basis in research. Vitamin E treatment has been shown to delay the onset of symptoms in a mouse model of ALS, although it did not have an impact on survival. It has now become common to recommend 2,000 I.U. of vitamin E per day in combination with riluzole as a treatment for ALS. Milk or some other form of fat in the stomach greatly enhances the absorption of vitamin E.

Other holistic remedies have not been proven valuable; however, other approaches, such as therapeutic massage and visual imagery, may offer support, encouragement, and hope. None of us can live without hope, and people with ALS should be encouraged to share their exploration into alternative treatments with their physicians. The physician should serve as an advisor, steering patients away from harmful therapies and towards those that may help. For those who desire a holistic approach, it is important to find an ALS physician who is willing to communicate with alternative medicine providers so that your overall ALS

treatment can be coordinated and beneficial. In this way, you will feel free to ask for more conventional therapies when the need arises. Ultimately, it is your decision as to how your disease will be treated.

Terminal Management

At some point in the progression of ALS, you may become tired of the struggle to continue living. Many people with ALS ultimately reach a level where they feel they can no longer maintain a satisfactory quality of life. At this point, the emphasis shifts from extending life to making the remaining days more comfortable (see Chapter 12). Nursing agencies can arrange home visits when you find it too difficult to go to the clinic for periodic evaluation. Most people desire to be pain-free at this point in the disease; they want to be as alert as possible during the day and have a comfortable, uninterrupted night's sleep. They should be given whatever treatment is necessary to relieve suffering, even if it does shorten life. Antianxiety medications (lorazepam), sleeping pills (temazepam), and pain relievers (including morphine) should be available as needed. People with ALS sometimes may require anti-nausea medications (prochlorperazine), because both shortness of breath and the medications to relieve it may cause stomach upset. Most people are reassured by the knowledge that their death will be peaceful and without struggle, lack of air, or pain.

Quality of Life

Although no one would ever choose to have ALS, many people with the disease affirm that ALS has brought blessings as well as suffering. They report new appreciation of family, friends, the beauty of nature, and life itself. Many people find the capacity to enjoy life each and every day. Morrie Schwartz, who had ALS, wrote, "After you have wept and grieved for your physical losses, cherish the functions and life that you have left." As health care providers, we consider it a privilege to be included in this intimate celebration of living and witness how ordinary people summon extraordinary faith and courage to overcome the hardships imposed by ALS and continue to live full lives.

