

LIVING WITH ALS  
RESOURCE GUIDE

6



# Managing Symptoms of ALS





# MANAGING SYMPTOMS OF ALS

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**Living with ALS**  
**Managing Symptoms of ALS**

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## INTRODUCTION

ALS may cause many different changes to your functioning, however; not everyone will experience every change and not all changes occur at the same time. As the disease progresses and various functions may become affected, it is helpful to understand these changes so that you know what to expect and how to manage these new changes. Always speak with your physician or other healthcare provider prior to starting any medication or treatment.

*The ALS Association was always a few steps ahead of us and that was really helpful. We never got to the next step of the disease without being prepared in advance for what to expect. That made the big changes seem like they weren't so bad after all because we were prepared.*

Brenda Davenport, her father, Frank, has ALS  
(Contributed by The ALS Association MN/ND/SD Chapter)

What we will cover in this resource guide:

- Mouth and nose issues
- Bowel and bladder symptoms
- Air hunger and breathing difficulties
- Insomnia and frequent awakenings
- Nutritional issues
- Skin problems
- Muscle changes
- Pain and fatigue
- Mood and emotions
- Thinking and behavior changes
- Pseudobulbar affect (excessive crying or laughing)

*My goal is to have the highest quality of life for the longest period of time.*

Dawn (Contributed by The ALS Association Golden West Chapter)

## MOUTH AND NOSE: ISSUES AND TREATMENT OPTIONS

### Sialorrhea (Drooling)

Sialorrhea, commonly known as drooling, is a frequent challenge with ALS. The medical term **sialorrhea** can be broken down into “**sial**,” meaning saliva and “**rrhea**,” meaning to flow. Normally we produce approximately 1.5 liters of saliva per day (Dand and Sakel 2010). Saliva does have an important purpose: It serves as a lubricant to keep the mouth clean and help with swallowing solids.

The excess buildup of saliva in a person with ALS is not usually due to an increase in the amount of saliva produced, but more often it is related to difficulty swallowing. Although appearing trivial in comparison to other ALS challenges, drooling can be a significant source of psychological distress and discomfort.

Swallowing requires coordination among the brain and nerves that control the movements of the mouth (lips, tongue) and the swallowing muscles in the throat and neck. In ALS, there is difficulty with this coordination, which can cause sialorrhea (Lakraj, Moghimi, and Jabbari, 2013) because it is difficult to move saliva to the back of the mouth and swallow it reflexively. The saliva pools in the front of the mouth and leaks out.

## Treatment Options

The goal of treatment is to reduce, but not totally eliminate saliva. The possible management strategies of sialorrhea (drooling) include:

1. The “**chin tuck**” is a simple strategy for both management of saliva and while eating (Figure 1). This is done with proper positioning (Dand and Sakel 2010). Ideally, you **sit in an upright position** making sure your head is stabilized. Then **slightly tuck your chin downward during swallowing to prevent choking**. If you are slouched or in a reclined position while attempting to swallow, this could lead to food or saliva going into the airway, causing choking.

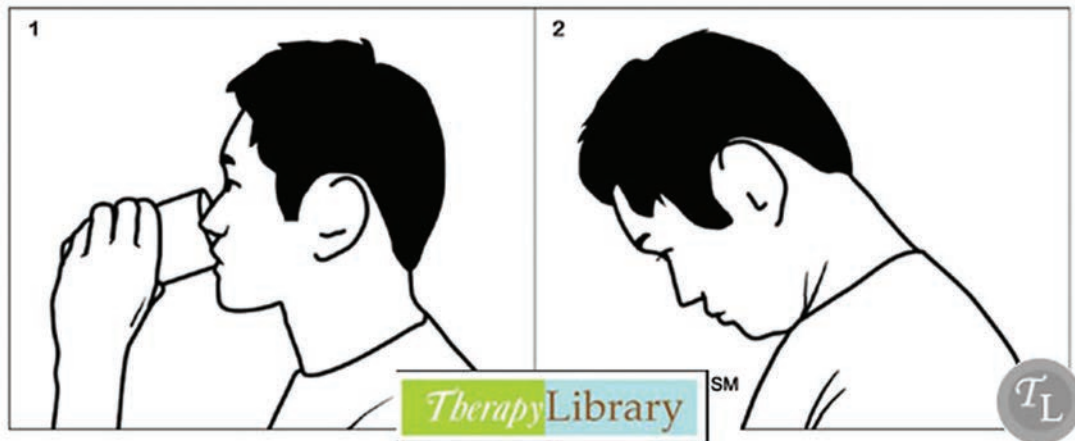


Figure 1: Example of the “chin tuck.” (Source: Therapy Library, 2009)

2. **Prescription medications** are a second option. These prescribed drugs **aim at decreasing the production of saliva**. Common medications include glycopyrrolate (pill), scopolamine (patch applied to the skin), amitriptyline (pill), or atropine (drops placed under the tongue). These medications can be used either alone or sometimes are combined to have a better effect.

Many healthcare providers prescribe **glycopyrrolate**, as it seems to have fewer side effects and is generally well tolerated. Scopolamine comes in a patch and is often prescribed when a person with ALS can no longer swallow.

The **scopolamine patch** lasts longer than glycopyrrolate, so you use one patch every three days.

**Amitriptyline** is an older medication also used to treat depression, but it is handy for a person with ALS because of its side effect of causing a dry mouth. **This medication can also make you sleepy.** If given at night, it helps improve difficulty sleeping (medical term: **insomnia**) and it dries up the saliva. Furthermore, it can help reduce involuntary crying and laughing (medical term: **pseudobulbar affect**).

**Atropine drops** are administered three or four times a day under the tongue. This medication has the advantage of a short duration of action.

The choice between medications is made based on how much drooling, when it occurs, and other problems that may need treatment (such as insomnia or pseudobulbar affect).

**If you experience sialorrhea that does not respond to medications, the next possible step would be to use injections such as Botulinum toxin (Botox).**

These injections are given into the salivary glands. They decrease the amount of saliva produced and can provide benefits for up to three months. The injections must be provided by a specialist and can be painful in their application.

3. If medications do not fully control your saliva, or if there are certain times of the day when saliva is worse, you can also use a **suction machine** to remove the saliva from your mouth. A suction machine that you use at home works like the suction device your dentist uses to clear saliva and rinse water from your mouth. It can be used as much or as little as you need throughout the day to pull saliva from your mouth.
4. **Radiation** can be applied to the salivary glands for the more severe cases. This will stop the gland from functioning. Care must be taken with this procedure as this may cause the once thin secretions to turn into a very thick “wallpaper paste” consistency. The drying effects from radiation on the salivary glands are permanent.
5. **Surgery (ablation)** to completely destroy the salivary glands has been done in Europe. This would be a last resort as it is irreversible and could potentially leave one with very thick secretions or an uncomfortably dry mouth.

## **Dry Mouth/Thick Secretions**

Normal breathing usually occurs through the nasal passages, which act to humidify the air before it reaches the lungs. The mouth in otherwise healthy persons is generally not used for breathing. In ALS, the muscles that normally keep the jaw and lips closed become weakened to the point that the mouth can no longer be closed and the mouth becomes the primary airway path of least resistance. This is called mouth breathing. **Mouth breathing causes the mouth to become dry, which results in thickening the saliva.**



Thickened secretions are more difficult to move and swallow. In a person with impaired swallowing or weakness of the mouth and tongue, the thickened saliva makes the problem worse.

### **Treatment Options: Increased Liquids or Use of Medications or Medical Devices**

1. **Liquids** help in thinning saliva and easing the movement of saliva towards the back of the mouth. **However, liquids are particularly difficult to swallow**, especially when swallowing abilities are poor. If a person is afraid of choking on liquids, he or she may intuitively drink less, but this leads to chronic dehydration. Chronic dehydration happens slowly over time and is often not recognized. The more dehydrated one becomes, the thicker the secretions.

**The treatment of thick secretions involves a combination of fluid management and sometimes the use of medications.** The first step is to avoid dehydration. According to Yorkston, Miller, and Strand (2004) the standard “liquid diet” or “blenderized” meal does not provide enough water in the system to prevent dehydration. People with ALS may have difficulties drinking thin liquids such as water, but may be able to drink something a bit thicker such as juice or thickened liquids. Gelatin, as well as ice pops, are other possible fluid options.

2. **Medications** are also used to manage thickened secretions. Medication choice is based upon the severity of the problem and what is causing the problem. Over-the-counter **guaifenesin** (an expectorant cough medicine) can be effective in thinning the saliva (Yorkston, Miller, and Strand, 2004).

**Papase** is an enzyme that can be obtained in supermarkets or drugstores. It is one of the ingredients in meat tenderizer and also found in papaya (juice). It has been reported to be beneficial in thinning secretions. Papase, when applied to the mouth, causes the thickened secretions to dissolve, allowing easier swallowing (Yorkston, Miller, and Strand, 2004).

3. A prescribed medication, called a “**mucoytic**,” can be used to thin secretions. These are often used when the thick secretions are very far back in the throat and you cannot seem to bring it forward to spit out, nor clear it with swallowing. **Acetylcysteine** is one of these mucoytic medications, given three times per day. **It is given using a machine called a nebulizer and the individual inhales the medication.** This medication is usually used for thicker secretions and when there is still sufficient coughing ability.
4. **Potassium iodide (SSKI)** can also be used. SSKI (a prescribed medication) acts as an irritant to the inside of the mouth, causing an increase in saliva. This will thin secretions. This SSKI treatment, however, may take up to two weeks to show effect (Yorkston, Miller, and Strand, 2004).
5. When fluids and medication are not enough for managing secretions, there are two types of medical devices that can help. **High Frequency Chest Wall Oscillation (HFCWO) devices**, originally used to thin secretions in people with cystic fibrosis, use an inflatable vest to vibrate the chest. The vest is connected to a machine, which uses an air compressor to quickly inflate and deflate the

vest. The air movement in and out of the vest creates vibrations, which loosen the bonds between mucous fibers in your lungs and airways. This helps to thin secretions, making them easier to cough up and swallow or suction out of your mouth.

6. Sometimes, even after secretions are thinned with fluids, medication, and/or a HFCWD, they can still be difficult to move or cough up. Weakness in your diaphragm (the muscle used most for breathing) and weakness in your throat make it hard to create enough pressure for a strong cough. To clear secretions in your throat and airways, you can try using a **cough assist device** (a mechanical insufflator-exsufflator). A cough assist device consists of a mask that connects, through tubing, to a small machine. The machine gently blows air (positive pressure) through the mask into your lungs, followed by pulling air (negative pressure) from your lungs. This simulates the pressure change that occurs during a natural cough and helps move secretions up and out of your airway.

## Excessive Yawning

Excessive yawning is experienced by a significant number of people with ALS. The cause is not well understood and, unfortunately, there are few effective treatments. A common antidepressant, Lexapro (escitalopram), lists yawning as a side effect. If you are taking this drug for depression and yawning has become a problem, you and your doctor may want to discuss changing the antidepressant medication.

### Treatment Options

Some people with ALS have reported relief of excessive yawning by sucking on hard candy or chewing gum. **Caution should be used if choking is a concern.**

## Jaw Quivering or Clenching

When you have ALS you may develop **muscle spasms** in any muscle. There are several muscles that control proper opening and closing of the jaw. **If the muscle spasm occurs in the muscles of the jaw, it can cause difficulty in either opening or closing of the mouth.** Jaw clenching can occur when the “jaw closers” or “jaw openers” develop a continuous and strong spasm. When this condition is severe, it can lead to jaw clenching, with biting of the sides of the tongue and cheeks (Elman and McCluskey, 2014). If this condition is sustained for a long period of time, it can produce a contracture (permanent and substantial shortening of the jaw muscles) (Clark, 2003).

### Treatment Options

1. Botulinum toxin (**Botox**) injections into the jaw-closing muscles can be used in cases of severe, sustained, jaw-closing spasm (Clark, 2003).

2. Medications that can relax muscles such as baclofen, tizanidine, and benzodiazepines can be used for cases of increased muscle tone (medical term: **spasticity**).
3. A referral can be made to a prosthodontist to consider jaw stretching exercises and dental appliances.

## Laryngospasm

Laryngospasm is a short-lived closure (most often lasting less than 30 seconds) of the voice box (medical term: **larynx**), which can be a terrifying experience. It is **caused by an involuntary and forceful closure (spasm) of the vocal cords**. This leads to a complete stop of airflow in and out of the lungs. It most often occurs in response to acid reflux or choking on food particles or liquids, including thickened saliva (Elman and McCluskey, 2014). It can also occur in response to irritants like perfumes, allergens, or smoke. During these episodes, people feel as though they cannot breathe. In severe cases, it can result in complete blockage of the upper airway (Kühnlein et al., 2008).

## Treatment Options

Immediate treatment during a laryngospasm episode includes **rapid repositioning of the head and neck to an upright position with the jaw forward**. This technique has been found to shorten these episodes (Kühnlein et al., 2008). You can also use breathing methods to shorten or abort the episodes. Try to breathe in slowly through your nose, and exhale forcefully through pursed lips (like you are breathing out through a straw) if you can. Understanding what laryngospasm is and that it rarely last more than seconds can help avoid a sense of panic that can worsen the laryngospasm. Long-term therapy would include lifestyle modifications, including smaller but more frequent meals and avoiding throat irritants. Medications such as short-acting benzodiazepines are used to help muscles relax. Medications to control acid reflux are also used to decrease the frequency of attacks (Sperfeld et al., 2005).

Although it happens very rarely, laryngospasms can be severe and make you feel lightheaded. When this happens, try to get into a seated position or lie down, if possible. If you are unable to move enough air in and out of your lungs during a laryngospasm, you may pass out (lose consciousness). While this is frightening, it is not life threatening. In fact, when you lose consciousness, the muscles throughout your body will relax, and the laryngospasm will stop. When the laryngospasm stops, you will be able to breathe again, and will quickly regain consciousness.

## Thrush

**Thrush is a fungal infection that can occasionally be seen in ALS.** It is often referred to as a “yeast infection.” The “candida” fungus is naturally found in the gastrointestinal and genitourinary tracts of all humans. They can invade, however,

and take up “residence” in a place that they are not normally found, causing an overgrowth.

When you suffer from dry mouth due to mouth breathing in ALS you are at a higher risk for developing this condition (Kauffman, 2013). Many people with thrush complain of a **“cottony” feeling in the mouth, decreased sense of taste, and, in some cases, pain when eating or swallowing.** **If you have thrush you may notice white plaque or patches** in the mouth and on the tongue, roof of the mouth (palate), and sometimes down into the throat.

### Treatment Options

The management of thrush involves an overall assessment and avoidance of risk factors. **Thrush is simply and effectively treated with application of antifungal medication** into the mouth (Singh et al., 2014).

### Nasal Congestion and Post-Nasal Drip

Any irritation or inflammation of the nose or the mucous membrane inside of the nose is referred to as **rhinitis**. Common symptoms include congestion, runny nose, sneezing, itching or obstruction, and post-nasal drip. There are two major forms of rhinitis, **allergic and non-allergic**. Allergic rhinitis occurs when an allergen triggers nasal symptoms. Non-allergic rhinitis occurs when there is no allergen, but irritation and inflammation of the nose occurs, caused by changes in the weather, strong odors, or cigarette smoke (Tran, Vickery, and Blaiss, 2011).

### Treatment Options

1. There are several treatments for rhinitis. The first is **avoidance of environmental triggers**. This can include strong odors from paints, perfumes, strong soaps as well as air pollutants such as smoke (including that from tobacco) that are known to be respiratory irritants (Tran, Vickery and Blaiss, 2011).
2. **Decongestants can help when used with intranasal corticosteroids, or topical antihistamines, or both.** Ipratropium bromide is a nasal spray recommended for a runny nose. Using this spray with a corticosteroid may be even more effective. People with mild symptoms often find relief with the use of a nasal saline spray. This can be effective to both soothe the nose if it feels dry and help to relieve nasal congestion.
3. **Antihistamines can also be used** for their drying properties. A common over-the-counter medication is Benadryl (diphenhydramine). It is important to read the label and use caution as these medications can be sedating.

Talk to your healthcare provider about what treatment is right for you.

## BOWELS AND BLADDER

### Urinary Urgency/Frequency

Most ALS information rarely focuses on issues of the bowel and bladder, leading people to believe the disease does not affect them. However, **urinary urgency (the “got to go” sensation), as well as constipation, may occur frequently** especially for people who experience more symptoms of muscle spasticity, which is an upper motor neuron symptom. Urinary urgency and constipation, combined with increased difficulties with mobility, can lead to discomfort and embarrassment.

Urinary urgency leads to “urge incontinence.” This is caused by over activity of the muscle in the wall of the bladder. Leakage of urine (incontinence) can occur before one has a chance to reach the toilet. **Some people try to cut back on fluid intake in an attempt to reduce leakage episodes. This can often make symptoms worse and cause dehydration.** The body will try to conserve water and the urine will become concentrated, which will also irritate the bladder lining (Griebing, 2009). Dehydration can also lead to constipation, which will be addressed later, and cause thickened secretions.

### Treatment Options

The treatment of choice depends upon the symptoms and severity. Individuals must play an active role in choosing a therapy that best fits one’s lifestyle.

**Treatments are either non-surgical or surgical as described below** (Griebing, 2009):

- 1. Non-surgical options:** Treatments include diet, scheduled toileting, pelvic floor exercises, protective undergarments, and medications.

**Diet:** Certain food and beverages make urinary symptoms worse because they act as a diuretic or as an irritant to the lining of the bladder. Caffeine, for example, is a diuretic. It is found in coffee, tea, chocolate, and some soda. Diuretics increase the need to urinate. Either eliminating caffeine or switching to a non-caffeinated product may improve the symptom. Alcohol and carbonated beverages irritate the bladder. Foods that are higher in acid or that contain large amounts of potassium can cause bladder irritation and urinary urgency and frequency.

**Scheduled toileting:** Effective in treating symptoms of urgency or urge incontinence when the bladder is full. Most people do not attempt to go to the bathroom unless they feel that the bladder is full. This, added to mobility difficulties, may result in not reaching the bathroom in time. Going to the toilet on a regular schedule to keep the bladder from becoming too full may be helpful.

**Pelvic floor exercise:** Commonly referred to as “Kegel” exercises, they strengthen the pelvic floor muscles, which support the bladder and rectum. These exercises can be done at any time by tightening, holding, and relaxing the pelvic floor muscles.

**Absorbent pads and products:** There are a wide variety of these products available on the market to help with incontinence. These are not a “cure” but are helpful with regard to managing the symptoms when you want to engage in physical or social activities. Men also have the option of wearing external male “condom” catheters. An adhesive condom attaches to tubing and a bag. Urine flows through the tube and into the bag, which can be attached to the leg under clothing. This not only catches urine leaks, but can be used to eliminate the need to travel to the bathroom throughout the day.

**Medication management:** There are several medications that work to suppress the urgency and also ensure effective urinary drainage. **Oxybutynin** is a medication that may help. It is taken by mouth up to three times a day. There is also an extended release version of this medication. Oxybutynin is also available in a patch that is absorbed through the skin (a transdermal patch) given two times per week. Tolterodine is another medication that is available in a long-acting form (Olek, 2014). Talk to your doctor about what might be best for you.

- 2. Surgical options:** Unlike the more common catheters that are inserted through the urethra (tube that allows urine to flow from the bladder to the outside), the suprapubic catheter is inserted through a hole or portal located just above the pubic bone. It drains the urine into a bag that can be attached to the leg (Boerner, 2010). The portal is created using a minor surgery that is commonly done as an outpatient procedure. The catheter is usually replaced on a monthly basis. This type of catheter has been reported to be more comfortable than the standard type of catheter placed in the urethra and is not associated with damage to the urethra due to insertion and removal procedures (Boerner, 2010). All catheters become colonized with bacteria over time, but treatment with antibiotics is only necessary if the person develops signs of infection.

## **Constipation/Diarrhea**

**Constipation** can be very distressing. **When treating this symptom we need to consider various causes, prevention, and as required, the use of medications such as a stool softener and a stimulant medication** (Andrews and Morgan, 2013). Lack of dietary fiber may be a common cause. There are several factors that can contribute to constipation specifically in ALS, such as lack of mobility. Increased weakness involving the abdominal muscles makes it more difficult to bear down and push the stool from the body. Some people may become constipated due to lack of fluid intake or related to medication side effects.

**Diarrhea is a less frequent problem;** however, many individuals with ALS will be started on liquid formulas to maintain weight when eating becomes problematic due to swallowing difficulties and fatigue. Many of these formulas contain increased amounts of fiber, producing loose stools (medical term: **laxative effect**).

## Treatment Options

1. One treatment option includes **increased fluid intake if dehydration** is believed to be contributing.
2. **Laxatives are often used for the treatment of constipation.** Laxatives can be separated into four main headings: softeners, stimulants, bulking agents, and osmotic agents.

**Stool softener:** Docusate sodium, dosage of up to 500 mg daily. This medication brings more fluid into the bowel. These can take several days to take effect.

**Stimulant laxatives:** Senna, bisacodyl, or sodium picosulfate. These medications have a stimulant effect. These can take effect in approximately 6-12 hours. Glycerol suppositories act as a mild irritant to the lining of the rectum. These can take effect in 15-60 minutes.

**Osmotic laxatives:** Magnesium salts and polyethylene glycol. These medications work to increase water absorption into the stool, making it softer, bulkier, and easier to pass.

3. **Non-medication management:** A recipe provided by the American Dietetic Association for constipation management, called **“black magic.”**
  - Three parts bran (wheat bran or 100% bran is best)
  - Two parts applesauce
  - One part prune juice

It is most effective if eaten three times per day. It can even be spread on toast.

If diarrhea occurs, treatment would involve assessment of the diet and possible dietary adjustment.

## RESPIRATORY SYMPTOMS

### Breathing Difficulties and “Air Hunger”

People with ALS easily recognize the weakness in the muscles in their arms and legs, but weakness in the muscles that are used for breathing (respiration) can be harder to recognize. The main muscle for breathing is called the **diaphragm**. Additional muscles in the chest wall support the breathing effort. When we breathe, we move air in and out of the chest. In the process, we take in oxygen and get rid of carbon dioxide through our lungs. If the respiratory muscles are not working correctly, the air exchange is impaired. As a result, the body sometimes does not get enough oxygen and there is a buildup of carbon dioxide.

**Weakness of the diaphragm often goes unnoticed when awake.** Sometimes, you may feel you cannot take a deep breath, especially when lying down flat, but more often than not, the symptoms caused by weak breathing muscles are very subtle. You may feel drowsy or foggy when you wake-up in the morning or feel sleepy all day. You may notice a headache first thing in the morning that quickly

disappears as the day goes on. These symptoms may indicate that you have a weak diaphragm muscle and do not have good breathing support during sleep.

Diaphragmatic muscle weakness is first evident when the body is in a sleep stage known as Rapid Eye Movement, or REM, sleep, because it is a state when we dream and most of the muscles in the body become completely limp. Normally, when a person is in REM sleep the diaphragm is the only respiratory muscle working (Barthlen, 1997). If there is weakness of the diaphragm, there will be impaired movement of air and buildup of carbon dioxide and lack of oxygen that can cause headaches or drowsiness upon waking. Weakness with moving air in and out of the lungs is called **hypoventilation** (not enough air exchange, or ventilation).

Sometimes people with ALS will also have difficulties with clearing secretions out of their airway due to a **weak cough** (Tripodoro and De Vito, 2008). This is also caused by muscle weakness and difficulty in coordinating the muscles of the throat, voice box, and belly.

According to the American Thoracic Society, **dyspnea is the term used to describe the sensation of breathing discomfort**. This discomfort varies in intensity. You may feel you cannot breathe when you lie flat (**orthopnea**). Some of the subtler symptoms are a feeling of restlessness or the inability to fall asleep or stay asleep. The body's response to decreased levels of oxygen (**hypoxemia**) is that it attempts to get more air into the body. If the body is not effectively able to get rid of the carbon dioxide, the level of this gas in the blood rises (**hypercapnia**). As the level of carbon dioxide rises, you may initially feel anxious. Over time, as this level remains high it works as an anesthetic and you can become sleepier.

With ALS, you may experience the sensation of very difficult or "labored" breathing. This symptom is also called "**air hunger**." People with ALS can develop labored breathing and air hunger in the final stages of ALS (Tripodoro and De Vito, 2008). These symptoms can be very effectively controlled with medication. You do not have to feel discomfort or anxiety from difficulty breathing.

## Treatment Options

The most effective treatment for weak breathing muscles is the use of a device to support breathing at night (nocturnal) while sleeping. Nocturnal **Noninvasive Ventilation (NIV) allows the respiratory muscles to be supported so that they can rest and recover at night**. This is "noninvasive" and involves the use of a mask. The mask can be taken on or off as desired.

One type of NIV, **Bilevel Positive Airway Pressure (BiPAP)**, is helpful to maintain an open airway. This type of support is called "bilevel" because it has two pressure levels, **one to breathe in (inhale) and the other to breathe out (exhale)**. BiPAP gives a higher pressure to "blow" air into the lungs and a lower pressure while you breathe out. The use of BiPAP support has allowed for better quality of life as well as longer survival in people with ALS (Barthlen, 1997).

Another type of NIV used for ALS is an **Average-Volume Assured Pressure (AVAP)** device. Like a BiPAP machine, it delivers higher pressures of air to help you inflate



your lungs, and lower levels of air while you exhale. The difference between the two devices is that a BiPAP device delivers set pressures, while an AVAP device delivers a certain volume of air (based on your body's needs), and will adjust the pressure to make sure the right volume is achieved. Because the pressure varies based on how much your own muscles are active in breathing, the pressure is generally lower while you are awake. Some people find this more comfortable and easier to get used to.

Some ventilators can be used in an AVAP setting and offer additional features such as a back-up battery (in the event that your power goes out), and a "sip and puff" option. "Sip and puff" refers to a second type of mouthpiece that can be used if you get short of breath with activities like talking or eating. Instead of using a mask (which is still used when sleeping or you need the support for longer periods), your machine can be switched to a straw-like connection. When you need help getting a full breath, you "sip" on the straw and the machine delivers a full breath of air. Many people with ALS like using this device to get full breaths between bites of food while eating, or words and sentences while talking. In order to use the sip and puff option (medical term: **open mouthpiece ventilation**), you need to have good facial strength and be able to purse and seal your lips around a large straw.

## INSOMNIA AND FREQUENT AWAKENING

### Multiple Reasons

Difficulties with falling asleep and with frequent awakenings are often secondary to the overall body changes associated with ALS. These can include anxiety and/or depression, the inability to change positions independently, or difficulties with breathing (such as dyspnea or orthopnea).

### Treatment Options

The first step is to identify the underlying cause. Often, underlying difficulties with respiration, as previously discussed, do cause difficulties with maintaining sleep.

**Sedatives are used with caution, as you do not want to decrease respirations further.** Some tricyclic antidepressants are used as they have a dual purpose of treating drooling or pseudobulbar affect and they have a side effect of making people sleepy.

## NUTRITIONAL ISSUES

ALS can make it difficult to take in enough food and liquids. Weakness in your jaw and tongue can affect your ability to chew and swallow safely. When swallowing becomes difficult, the first step is changing your diet to foods and liquids that are easier to swallow. Mealtimes often take longer, which contributes to feeling full and eating less. Some people with ALS have a feeling of excessive fullness when eating even small amounts (medical term: **early satiety**), or may feel full for many hours after eating (medical term: **delayed gastric emptying**). Others

may have a lack of appetite and not feel the need to eat as much as they did before the diagnosis of ALS. Muscle weakness in your arms and legs may make it hard to prepare meals or bring food to your mouth. All these changes can cause weight loss, malnutrition (not taking in enough food or nutrients to support body functions), and dehydration.

## Early Satiety and Lack of Appetite

**Loss of appetite is a common symptom in ALS. Feeling full before eating adequate amounts (early satiety) and feeling overly full for hours after eating because the stomach takes too long to empty (medical term: delayed gastric emptying) can lead to a decrease in appetite.** The cause is not well understood. There may be changes to the nerves in the gastrointestinal tract in persons with ALS that affect the movements that cause emptying of the stomach. Other symptoms caused by ALS, including constipation, lower levels of activity, depression, and weakness, can also contribute to early satiety and lack of appetite. Working with your physician or healthcare provider to identify the root cause or causes of your difficulty is the key to finding the best management strategies.

## Treatment Options

1. Sometimes taking too many over-the-counter herbal supplements can increase feelings of fullness and lower your appetite. You can try eliminating herbal supplements to see if your food intake improves.
2. If hand weakness and slowness of eating is problematic, make sure that you have enough help from your caregivers so that you can focus on chewing and swallowing rather than transporting food to your mouth.
3. Although mealtimes are important social events, you should not be expected to speak while chewing and swallowing. Have your caregiver explain to others that you need to spend your energy on eating rather than communication. Your tendency may be to isolate yourself at mealtimes; however, the social aspect of eating with others is important for both enjoying food and eating enough. We tend to eat more when we share our meals with others. Please note that having a feeding tube may alter this need to forgo communication during mealtime in favor of chewing and swallowing. We will talk more about feeding tubes in a later chapter of this manual.
4. Constipation and depression should also be addressed (see sections on bowels and bladder and mood/emotions).
5. Acid reflux is common in people with and without ALS, and can worsen early satiety and lack of appetite. Sometimes avoiding foods that trigger acid reflux is successful, but medications such as ranitidine (common brand Zantac), famotidine (common brand Pepcid), or omeprazole (common brand Nexium) can reduce acid and improve appetite.

6. If treating other causes does not improve appetite and food intake, several medications can be tried. Reglan (metoclopramide) speeds up the movement of food through the gastrointestinal tract and can reduce feelings of fullness or early satiety. For lack of appetite, medication options include mirtazapine (common brand Remeron) and dronabinol (common brand Marinol). Mirtazapine has the added benefit of helping depression and insomnia. In more extreme cases that have not responded to other treatments, a hormonal medication (Megace) is sometimes used. Medical marijuana may also be considered.

## Malnutrition/Dehydration

**Malnutrition occurs when the food and nutrients you take in from eating are not sufficient for the activities your body performs.** Calories from foods and liquids are your body's fuel. Liquids also moisten your tissues and are vital for your body's ability to eliminate waste. Not taking in enough calories will cause your body to use its own stores of fat and muscle to produce energy. While you may feel like losing fat stores will help you attain the body you've always dreamed of, loss of muscle can cause more weakness and speed up the progression of your disease. Without enough fuel, you will be more fatigued and may not be able to participate in activities you would otherwise enjoy. Protein, a specific source of calories, is vital for skin and cell repair. Lack of protein can make it harder to heal from injuries and contributes to skin breakdown (e.g., bedsores).

## Chewing and Swallowing Muscle Weakness

When weakness in your chewing and swallowing muscles makes eating more difficult, it becomes hard to take in enough calories and nutrients. Make the following changes to boost your calorie count:

- **Change the texture** of your foods to softer and moister foods that are easier to swallow.
- **Eat smaller amounts frequently** throughout the day.
- **Snack on high-calorie foods** or nutritional supplements (shakes like Ensure, Boost, and Carnation Instant Breakfast).
- **Have a speech-language pathologist evaluate your swallow** and recommend what foods will be easiest and safest for your particular weakness.
- **Increase your calorie intake by eating a high-calorie diet.** Use heavy cream instead of low-fat milk, add butter and sauces liberally, and avoid low-calorie options. A dietitian can provide you with lists of high-calorie food options that are safe for you, and offer recipes for high-calorie shakes, meals, and supplements. It is also important to take the "correct" types of calories specifically for you.

Difficulty with swallowing can also make it hard to drink enough to support your body's needs, and your body can become dehydrated. Limiting the amount that

you drink in order to prevent having to use the bathroom is another common cause of dehydration.

**Liquids help with many important body functions, including producing saliva, eliminating wastes, preventing constipation, and carrying energy and oxygen throughout your body. If trouble swallowing is causing you to become dehydrated, there are several steps you can take to increase your fluid intake:**

- **Drink thick liquids** like shakes instead of thin liquids like water.
- Specially designed powders and gels can be used to **thicken beverages** to make them easier to swallow.
- You can also add liquids to foods by using **sauces, cream, and gravy**.
- **Try sipping fluids throughout the day** rather than drinking large amounts at once.
- Your speech-language pathologist can give you strategies to help you swallow more effectively and efficiently, such as **tucking your chin, using a straw, or taking small sips**.

## Treatment Options

Eventually, ALS may weaken the chewing and swallowing muscles to the point where it is very tiring or even impossible to get enough nutrition and fluid orally (through your mouth). **Many people with ALS choose to have a feeding tube (enteral feeding) placed directly into their stomach to help supplement the food and liquid they are still able to eat and drink.** Sometimes drinking is more difficult than eating, and the tube is used for liquids but food is still eaten. Sometimes swallowing pills is a challenge, and the feeding tube is used for medications, but the person with ALS can still eat and drink sufficiently. Whatever the case, the feeding tube is an option to get nutrition, fluid, and medications that are difficult or impossible to swallow. Studies have shown that using a feeding tube can stabilize weight or even increase weight. **People with ALS often report that getting a feeding tube was a difficult decision, but most are glad they did.** Some frequent comments are that they wished they had the feeding tube much earlier, and that it greatly improved their quality of life.

**Having a feeding tube does not mean you need to stop eating or drinking by mouth.** Most people with ALS continue to eat and drink for pleasure as long as it is safe to do so. It also does not mean that your ALS will stop progressing. Although getting enough nutrition can prevent your body from breaking down its muscle for energy, your motor nerves will continue to be damaged, and weakness throughout your body will progress.

It is unclear whether feeding tubes significantly increase survival. Some studies indicate people with ALS who choose to use feeding tubes live longer than people with ALS who decline feeding tubes. However, other factors can influence the survival of these groups. It is likely that the better nutrition available to those with feeding tubes will improve their survival somewhat, but **the most compelling reason for using feeding tubes in ALS is for improving your quality of life.**

Perhaps you have already decided that getting a feeding tube is right for you. You may wonder when the best time to have the procedure is. **A feeding tube should be considered if:**

- You take a long time (over an hour) to eat
- You are fatigued after mealtimes
- You have been losing weight despite the use of a high-calorie diet
- You are spending a lot of your time or energy on taking in nutrition or liquids
- You have difficulty with swallowing food, liquids, or medications
- Your breathing number (Forced Vital Capacity or FVC) is getting close to 50%
- You are no longer able to socialize and enjoy meal times because you have to conserve energy for swallowing and breathing
- You know you want a feeding tube eventually

**Even if you don't have trouble with eating or drinking now, you may need to consider having a feeding tube placed proactively.** The benefit of doing this is that the procedure (minor surgery) is done while your breathing function is at its best. Studies indicate that there is a higher risk of complications, such as bleeding, infection, and pneumonia, to a certain type of feeding tube procedure if your **Forced Vital Capacity (FVC)** is below 50%. Forced vital capacity is the amount of air that can be forcibly exhaled from the lungs after taking in the deepest breath possible. This is used as a measure of lung disease or function. Your ALS clinic should monitor your FVC and talk about the option of having a feeding tube placed early, before your FVC drops below 50%.

**Regardless of whether or not you have a feeding tube placed, you can continue to eat foods that are safe for you to swallow.**

## SKIN PROBLEMS

Did you know that your skin is an organ? It serves multiple functions for the rest of your body. It protects your muscles, bones, and other organs. It shields against organisms like bacteria and viruses. It prevents water loss and temperature changes and gives you the sense of touch. **While ALS does not attack your skin, problems caused from ALS like not being able to change positions, lack of protein intake, and difficulty cleaning and drying your body can cause damage to your skin.**

### Fungal/Yeast Infection

Fungus/yeast is an organism that grows best in warm, dark, moist places. It usually infects areas where skin touches other skin (skin folds) like armpits, under breasts, the groin, inner thighs, and between fingers and toes. Keeping these areas clean and dry can both prevent and treat yeast infections. A yeast infection usually looks red and raw, and may be itchy or sore.



### Prevention/Treatment Options

- Keep skin clean and dry
- Expose skin folds to air
- Wear absorbent clothing (cotton and natural fibers)
- Over-the-counter antifungal creams can be used. If necessary, your provider may offer you a prescription strength antifungal.

### Bedsore

Bedsore are injuries to the skin caused from pressure or shearing (shifting between layers of the skin). Bedsore are more common in people with an inability to feel their skin (medical term: **sensory loss**), such as spinal cord injury patients, but can occur in anyone. They come in four stages, from mild redness and swelling (stage one) to loss of skin and muscle, all the way to the bone (stage four). Bedsore are relatively rare in ALS.

### Prevention/Treatment Options

- Use pressure-reducing cushions when sitting and mattresses when lying.
- Do not stay in the same position for long periods. Turn in bed and reposition or shift your weight when sitting in chairs.

- Keep skin clean and dry. Healthy skin is better able to withstand and repair damage from external forces.
- Avoid malnutrition and dehydration, which alter the skin's protective mechanisms and impair healing.
- Be sure to eat adequate protein, calories, and vitamin C, which are essential for cell repair.

## MUSCLE CHANGES

ALS is a disease of the motor neurons (the nerves that allow your brain to communicate with muscle fibers).

**When the communication through motor neurons is damaged, the normal activity in muscle fibers is disrupted.** If upper motor neurons (nerves from your brain to your spinal cord) are damaged, muscles can weaken, become tight (spastic), and be prone to muscle spasms. If lower motor neurons (nerves running from your spinal cord to muscles) are damaged, muscles become weak and show signs like atrophy (wasting away), fasciculations (twitches), and cramping (charley horses).

### Fasciculations

**Fasciculations, or muscle twitches, are brief contractions of the muscle fibers innervated by an individual lower motor neuron.** Everyone has occasional fasciculations, but they are more regularly seen in people with diseases affecting the lower motor neurons. If you tighten or use the muscle that is twitching, the fasciculation will stop, but can occur again when your muscle is back to rest. Many people do not notice their fasciculations until they have been pointed out. They are more noticeable during periods of rest such as when going to bed at night. Fasciculations are not painful, and generally, do not need treatment.

### Muscle Cramps

**Muscle cramps or charley horses involve a larger group of muscle fibers than a fasciculation, and rather than brief twitches, the muscle will contract or tighten for an extended duration.** Muscle cramps can cause severe pain and discomfort. Many people report that cramping is worse if they have overused muscles.



## Treatment Options

1. Specific movements that use specific muscles often trigger cramps. Avoiding such movements (when possible) is one technique to limit cramping. It can also be helpful to gradually stretch muscle groups before performing movements you find cause cramps or exercising the muscles involved.
2. You can try taking an over-the-counter calcium and magnesium supplement, since lack of calcium or magnesium can provoke muscle cramps.
3. **If cramping is frequent or bothersome, talk with your physician about strategies or medications to treat muscle cramps.** Some options include anticonvulsant (anti-seizure) medication (levetiracetam, gabapentin, and carbamazepine), muscle relaxers (baclofen and tizanidine), and benzodiazepines (clonazepam and diazepam). Medical marijuana may also be a consideration.

**You will need to weigh the risks and benefits of using medications, as many of these can cause sleepiness.** You also may be relying on the tightness of your muscles (medical term: **increased muscle tone** or **spasticity**) to help compensate for muscle weakness, so taking medications that reduce your muscle tone (tightness) might affect your ability to perform activities like standing and walking.

## Spasticity

**Spasticity refers to stiffness, tightness, or increased tone of muscles.** It occurs when upper motor nerves are damaged. Not everyone with ALS will have spasticity; those with upper motor nerve predominate ALS are much more likely to have spasticity that requires treatment than those with lower motor nerve predominant ALS.

**Spasticity can make movements slow and more difficult, and cause you to feel more fatigue.** Because spasticity involves an inability for muscles to relax, it can sometimes be helpful, such as when a person with ALS has leg weakness but is still able to stand due to muscle tightness from spasticity.

## Treatment Options

1. Being inactive for a long period of time can worsen spasticity. Many people with ALS find that spasticity is worse in the morning upon waking. Doing **gentle range of motion and stretching exercises** can loosen your muscles and make it easier to move.
2. **Your specific weakness, function (ability to do activities), and amount of spasticity should be considered before taking medication for spasticity.** If you and your doctor decide an anti-spasticity medication is right for you, there are several that can be tried. Some medication options include baclofen, tizanidine, diazepam, clonazepam, and dantrolene. **Baclofen and tizanidine, having fewer side effects and requiring less monitoring, are usually the first medications tried.** You should start with a low dose and increase it slowly



over time. This allows you to watch for side effects like sleepiness, dry mouth, and weakness. If one medication causes side effects, don't be afraid to try a different medication, as side effects vary from person to person and from medication to medication. Medical marijuana may also be a consideration.

3. **When spasticity occurs in a specific area, localized treatment with Botox injections can be used to relax the muscles involved.** This will increase weakness to the muscles and has the risk of spreading to nearby muscles.
4. If muscle and joint movements are limited because of spasticity or weakness, tendons and ligaments will tighten and shorten, causing contractures. Braces or splints may be needed to straighten body areas and prevent or minimize contractures.

## PAIN AND FATIGUE

### Muscle Fatigue

Muscle changes like weakness, tightness, and cramps make everyday activities more difficult. **You may have to work harder and it may take you longer to do activities that you used to think were easy.** You might need to use two hands instead of one to raise a glass, or lift your thighs higher to prevent tripping over your toes. All of this extra work and muscle use can leave you feeling tired and worn out. You may not move your body the same way you were previously able to move it, and muscles and joints may start to hurt. Adjusting to your body's new limitations can help you prevent pain and enjoy life to the fullest.

**Fatigue is common in ALS.** Picture your body as a vehicle trying to drive up a hill. If you take away some of the vehicle's horsepower (muscle strength) and add a brisk wind (spasticity), it's going to be harder and take longer to reach the top. Your body is like a machine that is working harder and harder just to get through normal activities. Take care of your body by giving it rest, good fuel (food and liquids), and not asking it to do more than it should.

### Energy Conservation to Prevent Fatigue

In addition to getting good rest and nutrition, there are many things that you can do to help prevent getting worn out and live a full life despite fatiguing more easily. **Energy conservation is a principle where you look at your activities and plan ways to use your energy for the most important tasks and in the most efficient way.** This lets you have energy for the things you most want to do, and allows you to spend the least amount of energy on other tasks. Energy is like a battery charge: You have a limited amount stored and need to decide what to spend it on. Some energy conservation techniques that you can try are:

- Use **adaptive equipment** (reachers, built-up utensils, lightweight glasses) to reduce your work.
- **Allow family and friends to help** with activities that are getting difficult.
- **Consider using a volunteer or hiring help** for household duties.

- Plan your day to **have rest periods** between scheduled activities.
- **Reduce or eliminate activities that do not contribute to your goals** or enjoyment of life.
- Consider using a **hiring agency or contract help**, if appropriate and affordable.

Using energy conservation techniques will let you reserve energy to do activities that are most important to you. You can learn in greater detail about energy conservation in the resource guide, *Functioning When Your Mobility is Affected by ALS*.

Fatigue can also be a side effect of medications that are used in ALS. It is a rare side effect of riluzole (brand name: Rilutek), but if you began to experience fatigue around the time that you started taking riluzole, try taking a “drug holiday” by stopping riluzole use for two weeks. If your fatigue improves, you will need to decide if taking riluzole is worth the fatigue it causes you. First talk with your healthcare provider about this option.

## Pain

**While the loss of motor neurons and muscle itself is not painful, problems that can develop from weakness, muscle tightness, or cramping, can cause pain.** Joints that are not properly stretched will tighten and ache. Old injuries causing back pain, neck pain, or joint pain may worsen due to loss of muscle support. New areas of injury can occur if you push your body to do activities that require more strength than your body has.

Like many things in life, the saying “an ounce of prevention is worth a pound of cure” applies here. **Be kind to your body; don’t work it beyond what it can handle.** Use good body mechanics to prevent injuries and worsening old injuries. Make sure your body is positioned well when resting and avoid falls. When you are no longer able to move joints (shoulders, elbows, wrists, fingers, knees, and ankles) through their full range of motion, have a caregiver perform gentle stretching exercises on your body regularly.

## Treatment Options

1. If prevention is not enough and you find yourself experiencing pain, **use these tips at home to make yourself more comfortable:**
  - **If a new injury occurs, make sure you have it evaluated** for problems that require medical attention.
  - New injuries like sprains or strains should be treated with **RICE**: Rest, Ice, Compression, and Elevation. Avoid using the affected area, apply an ice pack, wrap the area with an ace or compression bandage, and elevate it to reduce swelling.
  - Older injuries or pain lasting more than one week can be treated with ice or heat packs (whichever works for your pain). **Massage and gentle stretches** help loosen tight muscles and reduce pain.

2. **Over-the-counter medications** like acetaminophen (brand name: Tylenol) or NSAIDs (Nonsteroidal Anti-Inflammatory Drugs) can be used to reduce pain and inflammation. Always consult your physician or healthcare provider before starting medications:

- **Ibuprofen** (brand name: Motrin): A typical over-the-counter dose is 400 mg every 4 to 6 hours, but doses up to 800 mg every 8 hours can be used. Avoid using ibuprofen for longer than two weeks as it can cause irritation to your stomach lining. It is best to take ibuprofen with food.
- **Naproxen sodium** (brand name: Aleve): A typical over-the-counter dose is 220 mg every 12 hours, but doses up to 440 mg every 12 hours can be used. Avoid using naproxen for longer than two weeks as it can cause irritation to your stomach lining. It is best to take naproxen with food.
- **Acetaminophen** (brand name: Tylenol): A typical over-the-counter dose is 700 mg every 4 to 6 hours (FDA), but doses up to 1000 mg every 6 hours can be used. Do not take more than 4000 mg every 24 hours, as liver damage and death can occur. Acetaminophen does not reduce inflammation, but does help with pain.

**Make sure to read the warnings on any over-the-counter medication you try.**

NSAIDs should not be used in people with bleeding or clotting disorders, or if you are taking certain medications such as warfarin. Acetaminophen should not be used if you have liver damage. If you need to use medication longer than two weeks for pain, talk with your physician or healthcare provider.

3. In some cases, Tylenol or NSAIDs are not enough to control pain. For mild to moderate pain, the next step is often to use tramadol (brand name: Ultram). Tramadol is a weak opioid that has fewer side effects than stronger opioid medications. If tramadol is not effective, ask your medical team about using opioid/narcotic medications. Although opioids have side effects, including sleepiness and constipation, they are the most effective medications for moderate to severe pain. Medical marijuana may also be a consideration.

**You do not have to live in pain.**

4. There is also a specific type of pain that can be treated with medications other than NSAIDs and opioids called **neuropathic pain**. This type of pain is usually caused by injury or damage to the sensory nerves. Although ALS does not typically cause damage to sensory nerves, people with ALS may have damage to sensory nerves from other causes. People with neuropathic pain describe it as feeling pins-and-needles, burning, or electrical pain. It can be treated with anticonvulsants (gabapentin and pregabalin), antidepressants (tricyclic antidepressants and selective norepinephrine reuptake inhibitors), and topical agents (capsaicin cream and lidocaine).

## MOOD AND EMOTIONS

When you were given the diagnosis of ALS, you may have felt numb or detached. Or you may have felt angry, fearful, or sad. We all experience a wide variety of emotions, and your emotions will likely be very affected by your diagnosis and

the changes in your body. Your emotional reaction to the diagnosis may vary from one day to the next. These fluctuations in mood are normal and expected. It is normal to feel distress and grief: not just about having a fatal illness, but about all of the little losses along the way. Grief is the emotional response to loss and can trigger emotions like denial, anger, and sadness.

*The key is to recognize that the disease will progress and to try to anticipate the next phase so that you can prepare properly for it. This approach is much easier said than done because of the emotional and psychological aspects of this kind of thinking.*

Judith Massey (Contributed by The ALS Association Northern Ohio Chapter)

## Depression

Although sadness is a normal emotion that is often felt when grieving losses, **having a low mood that interferes with your ability to live and enjoy life is considered depression.** Depression is present in otherwise healthy individuals, but is more frequent in people facing a severe and debilitating illness. Many people never seek treatment for depressive symptoms, but the majority of people can get better with treatment. **Some common signs that you may be depressed include:**

- Low mood or sadness on most days of the week
- Feelings of guilt, hopelessness, or emptiness
- Loss of interest or pleasure in activities, including sex
- Difficulty sleeping or sleeping too much
- Overeating or under-eating
- Excessive crying
- Fatigue or tiredness
- Moving more slowly or quickly than usual
- Thoughts of worthlessness
- Difficulty thinking or concentrating
- Irritability, restlessness, or aggression
- Thoughts of hurting or killing yourself

**While some of the symptoms of depression are present in people with ALS due to the disease itself (such as slow movements due to spasticity, or under-eating due to swallowing difficulty), the presence of many depressive symptoms may indicate depression.** If the symptoms make it difficult for you to live your normal life or enjoy life, you should consider having treatment for depression.

## Treatment Options

1. Depression that is mild can often be treated with **psychotherapy or counseling**. There are two main types of psychotherapy: cognitive behavioral therapy and interpersonal therapy.

In **cognitive behavioral therapy**, a therapist helps you identify negative thinking and develop strategies to view challenging situations more clearly and respond to them more effectively. In **interpersonal therapy**, a therapist helps you to link your mood and disturbing life events in order to regain control of your mood and functioning. Therapy works best when you find a counselor with whom you feel comfortable. If your first experience with therapy was discouraging, try seeing a different therapist and talking through your goals and experiences at the first visit.

2. **There are also many medications that are successful in treating depression.**

The first line of medication treatment is usually a class of drugs called **SSRIs (Selective Serotonin Reuptake Inhibitors)**. These include such medications as sertraline (brand name: Zoloft), paroxetine (brand name: Paxil), citalopram (brand name: Celexa), and fluoxetine (brand name: Prozac). The most common side effects include sleepiness, nausea, and headaches, which usually resolve after one to two weeks. To limit these side effects, you can start with a half dose for the first week and increase to the full dose starting the second week of use.

**Like most medications, it is best to first start with the lowest dose that helps depression and increase slowly to the dose that is best for you.** You may not see improvement for up to four weeks, and the full effect of treatment may not occur until you have been on a specific dose for six to eight weeks. In some cases, depression can initially get worse, so have your family watch for signs of worsening depression or thoughts of suicide when starting on medication for depression.

3. **If the SSRI medications do not work for you or if you have side effects to multiple SSRI medications, other medication options should be considered. A newer class of medication called SNRIs (Serotonin and Norepinephrine Reuptake Inhibitors)** are not only helpful for depression, but are also being used for chronic pain and neuropathic pain. Duloxetine (brand name: Cymbalta) and venlafaxine (brand name: Effexor) are SNRIs. Other medications for depression that are not part of a specific class, but have their own novel mechanisms of action, can help with other symptoms such as decreased appetite (bupropion, brand name: Wellbutrin and mirtazapine, brand name: Remeron), insomnia (mirtazapine), and decreased sex drive (bupropion).
4. If one medication does not fully control your depression, a second medication can be added to augment the first. The medications usually used to augment are called atypical antipsychotics (generic names/common brand names: aripiprazole/Abilify, quetiapine/Seroquel, risperidone/Risperdal, olanzapine/Zyprexa, and lurasidone/Latuda).

5. For people with severe depression who want or try to harm themselves, medication and therapy may not be enough. For other treatment options speak with a mental health treatment professional.

**Whether you feel more comfortable trying psychotherapy or medication to treat your depression, the important thing is asking for help.** Living with untreated depression will make it harder to enjoy the time you have with family and friends, and will affect your ability to achieve your goals and dreams.

## Anxiety

**Anxiety is a feeling of worry, nervousness, or fear.** It can be a common response to new or stressful situations. Sometimes, anxiety can become overwhelming and interfere with normal activities or happiness. Some signs of anxiety include:

- Feeling nervous, anxious, or on edge
- Not being able to stop or control worrying
- Worrying too much about different things
- Being so restless that it is hard to sit still
- Becoming easily annoyed or irritable
- Feeling afraid as if something awful might happen
- Increased fatigue and muscular tension
- Difficulty sleeping or relaxing
- Headaches and pain in the neck, shoulders, and back
- Increased blood pressure or fast heart rate

**While some of the symptoms of anxiety are present in people with ALS due to the disease itself (such as such as increased fatigue and muscle tension), the presence of several anxiety symptoms may indicate generalized anxiety.** If the symptoms make it difficult for you to live your normal life or enjoy life, you should consider having treatment for anxiety.

## Treatment Options

Treatment for anxiety includes **behavioral approaches and medication.**

1. Counseling with a cognitive-behavioral therapist can help you identify, understand, and change your thinking and behavior patterns. You'll learn skills and strategies to view your situation more clearly and respond to it more effectively.
2. **Relaxation techniques** can help you train your body to respond more calmly. Types of relaxation techniques include meditation, adaptive yoga, and acupuncture. **Mindfulness-Based Stress Reduction (MBSR)** is a program that uses principles from yoga and meditation to reduce stress and anxiety. It is based on the ancient practice of mindfulness and teaches being present in the moment, observing and experiencing without judgment, deep relaxation, and gentle movement. It can help you examine your reactions to life's stressors and recognize that you can choose how to respond. Most communities have classes

available in yoga, meditation, and MBSR. There are also books, DVDs, and on-line resources.

3. In some cases, behavioral approaches may not be effective for controlling anxiety, and medications should be used. **The first treatment of choice is to use antidepressant medications (SSRIs), which are also proven effective for anxiety.** Examples of SSRIs used to treat anxiety are citalopram (brand name: Celexa), paroxetine (brand name: Paxil), and sertraline (brand name: Zoloft). The most common side effects include sleepiness, nausea, and headaches, which usually resolve after one to two weeks. To limit these side effects, you can start with a half dose for the first week, and increase to the full dose starting the second week of use. Like most medications, it is best to start with the lowest dose that helps anxiety first, and increase slowly to the dose that is best for you. You may not see improvement for up to four weeks, and the full effect of treatment may not occur until you have been on a specific dose for six to eight weeks.
4. **For short-term treatment of anxiety, low dose benzodiazepines (lorazepam, clonazepam, and diazepam) can be used.** These are most often used during the initial treatment with SSRIs until the SSRI takes effect. They can also be used for long-term therapy if antidepressants are not effective or cause intolerable side effects. Side effects include sleepiness, cognitive impairment, and weakness. When used long-term, these medications should not be stopped abruptly.

Just like depression, anxiety can impact your ability to enjoy life. Taking control of your anxiety can help you live life to the fullest.

## THINKING AND BEHAVIOR CHANGES

For a long time, it was thought that ALS did not involve changes to thinking or behavior. More recently, we have developed a better understanding that cognitive changes can occur with ALS. **Approximately half of all people with ALS may have changes in their cognitive ability, though most often the changes are mild.** Cognitive and behavioral changes with ALS fall into three main categories:

1. **ALS with cognitive impairment** refers to changes in areas of attention, cognitive flexibility, and word generation. Memory and the ability to understand the relationships of objects in space (medical term: **visuospatial function**) are generally unchanged.
2. **ALS with behavioral impairment** is when ALS is accompanied by changes in social interactions and behavior.
3. **ALS with dementia** is when the person with ALS acts 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. It may include altered social interaction, emotional blunting, loss of insight, language changes, inappropriate behavior, personality changes, emotional apathy, lack of empathy, dietary changes, or obsessive behaviors. Memory loss may not be present, but a person with ALS may still have dementia. Different diseases can cause dementia. We now know that ALS can, but does not always,

result in dementia. The type of thinking and behavior impairment observed in ALS is often different than the rapid forgetting that marks the onset of Alzheimer's type dementia.

**Although opinion varies on the best tests to use to diagnose thinking and behavior changes in ALS, it is generally agreed that people with ALS and their caregivers should be asked about changes in thinking, personality, and behavior.**

There are screening tests that can be performed during a regular visit to the clinic and you may be referred for more formal testing by a neuropsychologist. If changes are identified, decisions about future care should be discussed early in the disease, while thinking and decision-making abilities are at their best. While everyone with ALS should talk with their families and caregivers about their wishes, and put their wishes in writing by creating an Advance Healthcare Directive, it becomes even more important if you have cognitive impairment.

**Unfortunately, there is no available treatment for ALS with cognitive and behavioral changes.** If there are changes in thinking, personality, or behavior, **the best approaches are to adjust the environment to promote safety and lessen the impact on both the person with ALS and others.** Establishing a routine and avoiding distressing or risky situations can help to lessen the severity of behaviors. It may be embarrassing or upsetting to be around people who do not understand the impact of ALS on thinking and behavior. It is important for caregivers to have support and practice self-care away from the loved one with ALS to avoid caregiver burnout. Caring for a person with ALS and cognitive or behavior changes may require a larger team of caregivers, volunteers, and community services.

## **PSEUDOBULBAR AFFECT: EXCESSIVE CRYING AND/OR LAUGHING**

Some people with ALS experience excessive crying and/or laughing, also known as **pseudobulbar affect, emotional lability, or emotional incontinence.** It is caused by damage to specific tracts of nerves in the brain.

### **What Happens**

**Pseudobulbar affect** is uncontrollable, involuntary, sudden, and often frequent crying or laughing that can be unrelated to your mood, or excessive for the situation. It can be unprovoked, or it can occur when you would normally feel sad or happy, but not necessarily enough to make you cry or laugh. It can affect close to 50% of people with ALS, and can range from mild symptoms that do not require treatment, to severe symptoms impacting daily life. **It tends to be more common in the bulbar form of ALS, which affects speech and swallowing.**

Sometimes laughing or crying occurs at inappropriate times, such as laughing during a funeral or crying when a joke is told. It can lead to frustration, humiliation, embarrassment, social phobia, withdrawal, isolation, and caregiver distress. Because it can have an impact on relationships and quality of life, it is important to consider treating pseudobulbar affect when symptoms become bothersome.



## Treatment Options

Several treatment options are available to address excessive crying or laughing:

1. **Nuedexta** (dextromethorphan-quinidine) is approved specifically for the treatment of pseudobulbar affect. It is generally well tolerated, with very few side effects.
2. **SSRIs** (sertraline, fluvoxamine, fluoxetine) have been used for pseudobulbar affect for many years (prior to the use of Nuedexta). Because they are also used for depression and anxiety, individuals with these conditions can take one medication to treat more than one symptom.
3. **Tricyclic antidepressants** (amitriptyline, nortriptyline) have also been used for pseudobulbar affect prior to the use of Nuedexta. In addition to treating pseudobulbar affect, they are also helpful for reducing saliva and treating insomnia. If you have more than one of these symptoms, you may want to use a tricyclic antidepressant to treat more than one problem.

If one medication does not help your crying or laughing, don't be afraid to ask your physician to try a different medication. **Gaining control over your pseudobulbar affect can have a huge impact on your quality of life.**

## SUMMARY STATEMENT

ALS is a neurological disorder that results in damage to motor neurons and primarily muscle function. **ALS, however, also has an impact on many different body functions, emotions, thinking and behavior, and the ability to function in everyday life.** For example, not being able to easily bathe and move can cause skin sores, and not being able to chew and swallow can lead to malnutrition and dehydration. All people with ALS will not experience every possible symptom discussed. The good news is there are ways to prevent and treat the symptoms associated with ALS. It is good to know in advance all the ways ALS may affect you so you can recognize when you need to make changes to prevent or treat symptoms to maximize your comfort.

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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](http://www.alsa.org).