LIVING WITH ALS 8

RESOURCE GUIDE

Adjusting to Swallowing Changes and Nutritional Management in ALS

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ADJUSTING TO SWALLOWING CHANGES AND NUTRITIONAL MANAGEMENT IN ALS

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Living With ALS
Adjusting to Swallowing Changes and Nutritional Management in ALS

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INTRODUCTION

The muscles involved in swallowing can be affected by ALS just like muscles used for mobility and breathing. Your health and nutrition are affected by your ability to swallow foods and beverages. Because swallowing and nutrition are related, two healthcare professionals, namely a **Speech-Language Pathologist (SLP)** and your **Registered Dietitian/Nutritionist (RDN**) typically work together to maintain your most functional swallow and nutrition status.

This resource guide will help you understand how swallowing is affected in ALS and what you can do to maintain nutrition for energy and strength and to keep your airway open.

What we will cover in this resource guide:

- Normal swallowing processes
- The role of the speech-language pathologist
- How ALS affects swallowing
- Assessment of swallowing problems (dysphagia)
- Optimizing diet with impaired swallow
- Maintaining hydration and nutrition status
- The role of the registered dietitian
- Safe swallowing strategies
- Educational resources for safe swallowing
- The option of feeding tube placement

UNDERSTANDING SWALLOWING

The Normal Swallowing Process

The primary purpose of swallowing is to safely transport food and liquid from the mouth to the stomach. **The act of swallowing is a complex process that involves approximately 26 pairs of muscles and five cranial nerves that work together** to propel food from your mouth to the stomach.

The swallowing process has been divided into four conceptual stages, namely the **oral preparatory, oral, pharyngeal,** and **esophageal stages**. Typically, food and liquid pass from our mouth and through our throat in less than two seconds, however, the sequence of events is very involved. The following events occur during each stage:

Oral Preparatory Stage: Food is placed into the mouth either with a feeding utensil (fork or spoon), fingers, cup, or straw, and then is chewed and prepared for swallowing. This typically involves a hand-to-mouth transfer motion as food is taken from a plate and placed into the mouth, or when a cup is picked up and placed at the lips for drinking.

Oral Stage: The muscles of the mouth, jaw, and tongue work to prepare the food material for swallowing. This involves chewing and grinding food materials into smaller participles. During chewing and grinding of food materials, your saliva

mixes with the ingested material to aid in forming a cohesive "bolus" that can be easily swallowed. Once the bolus is formed, muscles of the tongue work to push the food/liquid from the front to the back of the mouth and towards the throat. At this time, it is important for the lips to tightly close or seal off the mouth and for the muscles of the roof of the mouth (soft palate) to close off the entrance to the nose (nasal cavity). This ensures that food and liquid are directed into the throat and ensure that it does not escape out of the mouth or the nasal cavity.

Pharyngeal Stage: Once the ingested materials pass the tonsils, the pharyngeal stage of swallowing begins and the swallowing process becomes involuntary (not under your conscious control). The cylindrical throat muscles squeeze to help push the food from the upper throat, through a ring muscle at the bottom of the throat surrounding the opening of the food pipe (medical term: **sphincter**), into the food pipe (medical term: **esophagus**).

During this stage of swallowing, entrance to the airway (medical term: **trachea**) is protected by a flap of cartilage called the epiglottis. This flips over (down) during the swallow to route food/liquid away from the airway and towards the stomach and has often been referred to as the "guardian angel" of the airway. Two additional barriers occur during this stage to protect your airway, consisting of the closure of both the false and true vocal folds at the entrance to your trachea or airway.

Esophageal Stage: The ring muscle that divides the throat from the entry to the food pipe (medical term: **Upper Esophageal Sphincter or UES**) is usually closed to prevent 1) air from entering the stomach and 2) previously ingested food and liquid materials from coming back up into the throat (reflux or regurgitation). This sphincter briefly opens or relaxes during the swallow and at the start of the esophageal stage to allow both food and liquid to enter the food pipe (**Figure 1**). Once food or liquids enter the esophagus, or food pipe, a contraction of the muscle helps to move the food from the top to the bottom of the pipe (21-27 cm in length) and into the stomach.

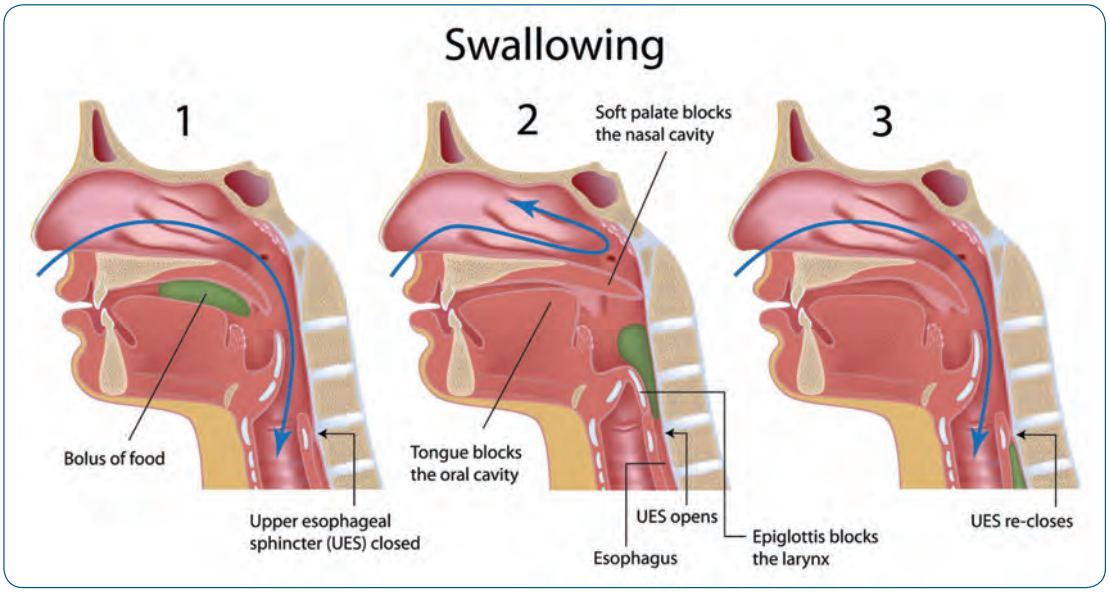

Figure 1: Swallowing process.

Swallowing Impairment

Dysphagia (pronounced dis-fay-ja) and Its Causes

Dysphagia is the medical term for a **swallowing disorder** or swallowing impairment during any of the four stages of swallowing. Swallowing impairment, or dysphagia, can occur in any stage of swallowing, and often occurs in more than one stage. **Dysphagia** is a symptom or consequence of an underlying disease, such as ALS.

Dysphagia is characterized in terms of the type and severity of difficulty during any of the four "stages" of swallowing. Some individuals will experience difficulties only during the oral stage of swallowing, while others will demonstrate impairments later in the swallow during the pharyngeal or esophageal stages. Some people will experience difficulties across all stages. Management strategies are appropriately prescribed to target the specific type of swallowing impairments you experience.

When treating dysphagia, it is important to evaluate the extent of the problem. That is, during which stage are swallowing difficulties experienced? Did entry of food into the windpipe instead of the food pipe (medical term: **aspiration**) occur? Was a strong and effective cough reflex triggered? Is there food or liquid left in the mouth or throat after the swallow occurs? What can be done to prevent or lessen these difficulties? These questions can be answered in a **swallowing evaluation**, as described later in this resource guide.

One important aspect of swallowing is protecting the windpipe or airway during swallowing. When airway protection does not occur, material enters the airway (medical term: **trachea**) and this is called **aspiration**.

Aspiration is a serious health concern because ingested material that made it into the airway collects bacteria and can often lead to lung infection and so-called **aspiration pneumonia**. Healthy individuals will elicit a strong cough reflex when food or liquids enter the windpipe to protect the airway and eject any materials; however, in some individuals with dysphagia, entry of food or liquids into the airway is not sensed (no cough or throat clearing occurs to eject the material out of the airway). This is called **silent aspiration**.

Although they can happen at the same time, it is important to distinguish a *swallowing* disorder from a *feeding* disorder in order to decide on the best plan of management. A feeding disorder involves difficulty transferring the meal from the plate to the mouth (e.g., due to arm, hand, or finger weakness) and can often occur at the same time as swallowing disorders. Your Occupational Therapist (OT) can provide adaptive utensils, meal trays, and feeding strategies to further assist with feeding-related impairments.

Dysphagia can occur due to many different disease processes or conditions that include, but are not limited to congenital disorders such as cleft lip and palate, cancer of the head, neck, or esophageal structures, stroke, Parkinson's Disease (PD), Multiple Sclerosis (MS) and ALS.

People with ALS may experience 1) swallowing difficulty due to the weakness and/or rigidity of the swallowing muscles and 2) difficulties with protecting the airway during swallowing.

HOW DOES ALS AFFECT SWALLOWING?

The ALS disease process involves degeneration of both Upper Motor Neurons (UMN) and Lower Motor Neurons (LMN). **Upper and lower motor neuron degeneration affecting head and neck muscles leads to increased swallowing difficulty and speech that is more difficult to produce and understand.** It also causes muscle weakness and stiffness of the **expiratory muscles** (breathing muscles) that further contributes to impaired airway protection during swallowing as you may not be able to generate enough force when breathing out (expiratory pressure) to produce a productive and effective cough (**Figure 2**).

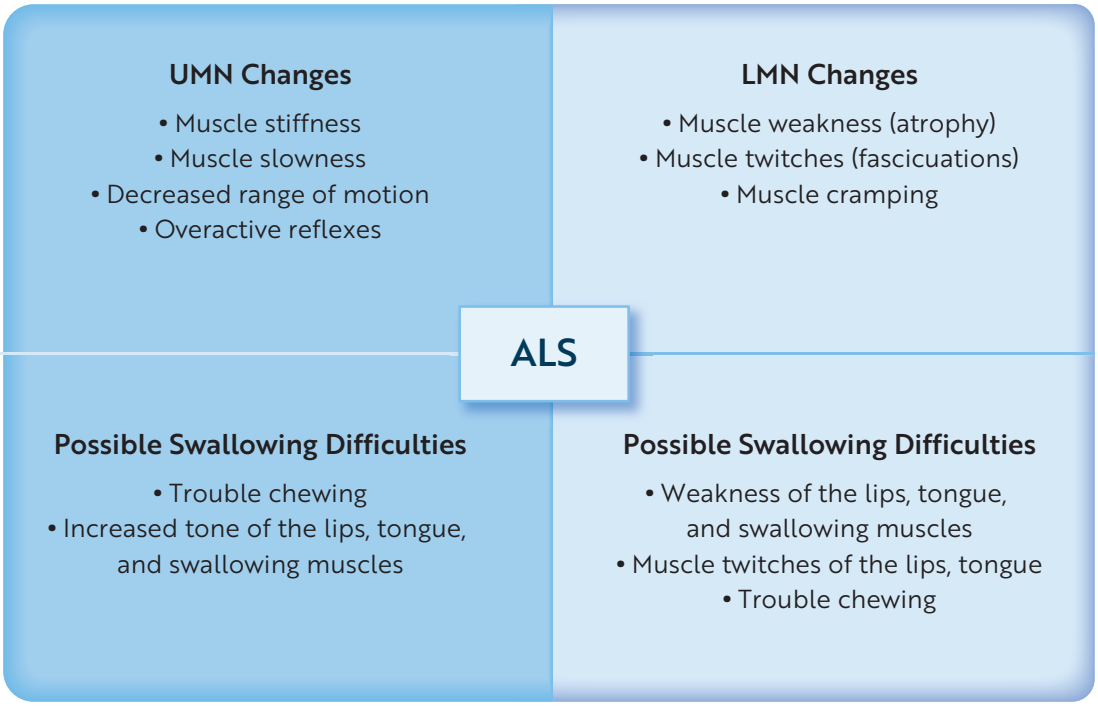

Figure 2: The symptoms of ALS divided by upper and lower motor neuron changes, with possible associated swallowing-related difficulties.

How Common Is Swallowing Impairment in ALS?

Approximately **85% of people living with ALS will experience dysphagia** at some point during the disease (Carpenter, McDonald and Howard, 1978; Chen and Garrett, 2005). This typically occurs during later stages of the disease; however, individuals who have a **"bulbar onset" (ALS that starts in the speech or swallowing muscles)** will likely experience swallowing-related difficulties much earlier during the disease.

It is important to be aware of possible difficulties in swallowing and red flags for unsafe swallowing so your healthcare professionals can help you make changes to keep swallowing safe.

Typical Swallowing Difficulties When You Have ALS

Swallowing difficulty can occur during any stage of the swallowing process. You may experience one or several symptoms we discuss; however, it is important to be aware of what may develop so appropriate changes can be made to ensure safe swallowing. You may begin experiencing symptoms and find they progress quickly. For this reason, **frequent monitoring by your SLP will help to identify and intervene** in order to keep swallowing safe.

1. During the first stage of swallowing (medical term: **oral stage**), you may experience a "**heavy tongue**" that makes it difficult to move and control food and liquid inside your mouth, chew, and clear all material from your mouth once you swallow. Due to lip weakness, food or liquid may spill out the front of your mouth while eating.
2. Impairments in the second stage of swallowing (medical term: **pharyngeal stage**) may include a **sensation of food "sticking" in your throat** after you swallow, **coughing frequently during the meal, or food/liquid coming out your nose**.
3. Regurgitation of food or liquid during or after the meal may be an indication of problems with the third stage of swallowing (medical term: **esophageal stage**). The onset of swallowing difficulty varies, so being aware of potential difficulties is beneficial. If you are experiencing any of these symptoms, it is important that your SLP and neurologist are aware so they can make appropriate recommendations. These difficulties and other swallowing-related impairments are listed in **Table 1**.

Table 1: Typical Swallowing Difficulties in People With ALS

Stage of Swallowing	Swallowing Difficulty
Oral Preparatory/Oral Stage	• Difficulty managing saliva • Difficulty chewing/fatigue with chewing • Food/liquid spilling out through the lips • Drooling • Difficulty controlling food/liquid in the mouth • Difficulty pushing the food/liquid to the back of the mouth • Residue in the mouth and cheek
Pharyngeal Stage	• Food "sticking" in the throat • Food/liquid coming out the nose • Coughing or choking during mealtime • Shortness of breath and fatigue during mealtime • Reduced cough strength and effectiveness
Esophageal Stage	• Regurgitation of food/liquid into the throat and mouth • Food "sticking" in the throat/base of neck

Risk of Aspiration and Malnutrition When You Have ALS

Many people with ALS have entry of saliva or food into their airway without noticing it happening. This is called **silent aspiration** and is something about which you need to be very aware. Therefore, it is very important to closely

evaluate swallowing function with the use of the **Modified Barium Swallowing study**, which is the gold standard test for identifying swallowing impairment and airway invasion. During this test, you are asked to swallow different consistencies of liquids and food while the swallowing process is filmed with an X-ray machine. More details about this test are given below.

HOW IS SWALLOWING EVALUATED?

The Role of the Speech-Language Pathologist (SLP)

The role of the SLP is to monitor both swallowing function and speech and to identify changes in function throughout the disease process. Once the SLP identifies a change or decline in swallow function, he or she will make specific recommendations to ease swallowing and mealtimes, and compensate for current difficulties. These recommendations may include diet modifications, changes in posture during swallowing, and safe swallowing strategies, all of which will be outlined in this resource guide. Recommendations and mealtime modifications will be tailored to you, based on your specific swallowing difficulty.

The Swallowing Evaluation Process

A comprehensive evaluation of swallowing **includes both a clinical "bedside" examination and an "instrumental" swallowing assessment**.

The bedside exam: Includes a series of questions focused on determining what difficulties you may be experiencing, a physical exam of the muscles of your face and mouth, and observations of different swallowing tasks (e.g., drinking water or chewing a cracker). This part of the exam helps your **SLP** develop a potential cause of your swallowing impairment, determine questions to answer during the instrumental assessment, and set up a possible treatment plan.

Modified Barium Swallow (MBS) study: This is the "gold standard" instrumental examination of swallowing. This is often referred to as a **swallow study** or **MBS** and involves looking at your dynamic swallowing function with a **real-time video X-ray** recorded and analyzed and typically played back to you. An SLP and radiologist will perform this study within about **15 minutes**; however, radiation is only used for small windows of time during the actual swallowing tasks and is generally limited to less than three minutes. **During this exam, you will be asked to swallow a series of liquids and food materials so that the medical team can evaluate the specific timing and movement patterns of you swallowing various types of textures.** Barium sulfate, which is visible under X-ray, will be used during this exam.

The purpose of the MBS is to assess the ability of the different muscles involved in swallowing to safely and efficiently move the food or liquid from the lips to the stomach. In addition, it can determine the impact of various techniques (discussed later in this resource guide) that are effective at improving swallowing movements or airway protection during swallowing. In people who are at high risk for **silent aspiration** (material entering the airway with no cough response),

this exam is very useful for diagnosis and management recommendations. The results of the study help educate you and your family/caregivers about the swallowing process, and guide recommendations for the safest diet you can take by mouth, as well as strategies to make swallowing as safe as possible.

An additional tool used to evaluate swallowing is the **Fiberoptic Endoscopic Evaluation of Swallowing (FEES) examination**. This involves using a specialized small camera with a light source embedded at the end of the tubing. This tubing is gently passed through your nose and can sit in your throat to visualize the muscles and structures in the throat during swallowing.

SAFE SWALLOW STRATEGIES

Your SLP and Registered Dietician/Nutritionist (RDN) may recommend different strategies to help compensate for specific difficulties in swallowing while maintaining nutrition. The purpose of these strategies is to make mealtime easier, more manageable, and safe. Strategies commonly recommended for swallowing and mealtime, and why they may help, are outlined in **Table 2**.

Table 2: Common Swallowing Strategies Recommended by Your Speech-Language Pathologist

Swallow Strategy	Description	May Help...
Effortful Swallow	Swallowing *hard*, squeezing all of your throat muscles as hard as you can	Reduce or eliminate "leftovers" or residue in the throat
Chin Tuck Posture	Tucking your chin down to your chest while you swallow	Help protect food or liquid from entering your airway
Small Bites/Single Sips	Taking small, single bites of food and sips of liquid	Reduce exertion and fatigue during the meal
Double Swallow	Swallowing 2x per sip of liquid or bite of solid/soft/pureed food	Help to eliminate food/liquid left over after the initial swallow

Red Flags: Signs That Indicate Trouble Swallowing

Certain symptoms indicate difficulty swallowing and can be observed during mealtime. **Symptoms that indicate a possible swallowing impairment include:**

- Coughing and/or choking on food or liquid while swallowing
- A wet or gurgling-sounding voice immediately after swallowing food or liquid
- Difficulty chewing
- Food escaping out of the mouth during chewing or liquid spilling from the lips
- Increased mealtimes
- The need for smaller bites and/or sips
- Drooling of saliva or liquids
- Food coming out the nose
- Regurgitation

- Difficulty managing secretions (saliva) during meal and throughout the day
- Shortness of breath during mealtimes

DIETARY MODIFICATIONS

Modified Diets and Mealtime Compensations

The purpose of modifying the consistency of food or liquids is to compensate for swallowing difficulties you might be experiencing. **Altering the consistency to a more appropriate texture will help reduce energy expenditure during feeding, chewing, and swallowing.** This will allow you to conserve energy throughout the day and experience less fatigue during mealtimes. Eating foods that require minimal chewing and moistening foods with sauces and gravies helps to reduce mealtime fatigue and eating duration.

In the throat, there are two sets of naturally occurring "pockets" that can catch foods, especially if the muscles involved in swallowing are impaired or weakened. Oftentimes this causes a sensation of "food sticking" or residue in the throat. You may feel the need to swallow an additional time in order to pass the residue and alleviate the sensation of food sticking. **Moistening foods with sauces or gravies can serve as a lubricant and ease the passage of the food through your throat during swallowing and may reduce the likelihood that the materials will get stuck in the pockets in your throat.**

Taking smaller bites and sips of food and liquid respectively may make it easier to control the food during the swallow. Additionally, alternating a sip of liquid every one to two bites of food may help to push the food materials down to your stomach. If medications become difficult to swallow whole, most can be crushed and taken with a tablespoon of yogurt/pudding or provided in liquid form (consult your physician or pharmacist for verification). Four different levels of modified diets are reviewed in **Table 3**.

Usually, **individuals with safe swallowing do best eating a mechanical soft diet**. This requires less chewing during the oral preparatory and oral phases of swallowing. Some people, however, may require a more restricted diet if their swallowing is deemed unsafe.

Coughing, choking, or difficulty swallowing may occur even with the use of mealtime compensations and dietary modifications. At that time, it may be beneficial to undergo a **Modified Barium Swallow** study to determine the safest diet consistency and safe swallowing recommendations.

Using Thickeners

The purpose of thickening agents or thickeners is to make regular liquids thicker (more viscous) and slow the flow rate of the liquid material during swallowing. Recall that swallowing occurs in less than 2 seconds. During this time over 26 pairs of muscles and 5 different cranial nerves need to coordinate and move

Table 3: Dysphagia Diet Levels and Appropriate Foods to Eat

Food Diet Level	Examples of Food in This Level
Level 1: Pureed	• Pudding • Pureed oatmeal, breads, meats • Hummus • Pureed fruits and vegetables
Level 2: Mechanical Soft	• Scrambled eggs • Meatloaf • Well-cooked vegetables • Pancakes • Mashed potatoes • Canned/cooked soft fruits
Level 3: Advanced	• Bread slices • Muffins • Moistened cereals • Pasta, casseroles • Baked potatoes • Soft/ripe fruits • Fish
Level 4: Regular	No Food Avoidances or Restrictions

in a complex pattern to protect the airway and direct the ingested materials towards the esophagus (food pipe) and away from the windpipe.

Adding a recommended thickening agent to liquids allows the swallowing system greater time to coordinate and protect the airway because the thicker liquids move at a slower speed, and in some individuals allows greater control of liquid material during swallowing (**Table 4**).

Like the levels of solid food modification, **there are also different levels of thickening for liquids**. Thickeners come in powder (starch based) and gel form and can be added to many of your favorite drinks to make them safer and more manageable. Your SLP will provide you with information on how to thicken your fluids to the recommended consistency. Some examples of **brands of thickening agents** are, Nestle® Resource Thicken Up, Simply Thick, and Thick-It.

Coughing or throat clearing during or directly after drinking liquids is a sign of **aspiration**.

Table 4: Thickening Liquids

Nectar-Thickened Liquids	• Liquid is a consistency slightly thicker than water • Does not contain fruit nectar or nectar flavoring
Honey-Thickened Liquids	• Liquid resembles the consistency of honey at room temperature

Preparing Food for Recommended Diet Levels: Tips for Caregivers

- If your loved one is using thickened liquids, keep a large container of thickened fluid in the refrigerator for easy access and use throughout the day.

- Reinforce safe swallowing strategies recommended by your SLP (for example: small bites or single sips, alternating bites of solid food with sips of liquid, performing a chin tuck during swallowing, or an "effortful swallow").
- Use a blender to soften foods that require a lot of chewing (i.e., breads, meats, raw fruits, and vegetables).
- Use your resources (see below for links to websites and handouts on swallowing difficulty).
- To minimize fatigue during meals, prepare small, easy snacks throughout the day.

Mealtime Strategies to Maintain Nutrition

In order to maintain adequate nutrition, it is important to be aware of signs of change in mealtime behaviors and swallow function. **Table 5** displays possible symptoms and strategies to combat changes noticed at mealtime.

Table 5: Mealtime Strategies to Ease Swallowing Difficulty

Possible Changes at Mealtime	Recommended Compensation or Strategy*
Longer mealtime duration	• Take smaller, more frequent meals throughout the day • Add snacks
Difficulty chewing	• Moisten foods with gravies and/or sauces • Use smaller bite sizes • Change food consistency to a softer diet that requires less chewing
Weight loss	• Add foods high in calories (e.g., peanut butter added to shakes, butter, condiments) • Drink full cream milk smoothies or shakes • Supplement nutrition with drinks such as Boost®, Ensure®, or Resource Benecalorie® • Add snacks throughout the day
Loss of enjoyment	• Increase taste, temperature, and textures using spices/sauces
Fatigue/shortness of breath	• Take smaller, more frequent meals throughout the day • Minimize exertion during meals (i.e., side conversations, distractions)

* This list of compensations and strategies is provided as a general guide. Please consult your Speech-Language Pathologist to find out which specific strategies are most appropriate for you or your loved one. (Chart adapted from Yorkston, Miller, and Strand, 2003.)

To further combat the challenge of fatigue that comes with muscle weakness, work with your team's OT to optimize feeding setup. Simple measures, such as having the plate on the same plane as your mouth (think sitting in a regular chair height at a bar counter) can decrease the arm and shoulder fatigue of hand-to-mouth. You can also stack phone books or use a plastic tub to elevate your place setting. Your OT can coach you on products that increase your eating/drinking efficiency to decrease fatigue. Some useful tools are: plate guards to spare you from chasing food and using the most efficient scooping food-to-mouth method, modified utensils, etc.

MAXIMIZING NUTRITION WHEN YOU HAVE ALS

Malnutrition Risk in ALS

Due to the disease process, individuals with ALS are particularly at risk for malnutrition for two reasons:

1. The presence of **hypermetabolism** (when you burn calories faster than "normal") (Desport et al., 2001)
2. **Reduced caloric intake** because of eating less due to swallowing problems and fatigue (Ngo et al., 2014)

Simply put, when you have a higher resting metabolic rate and you need a higher number of calories to maintain balance, but you typically consume fewer calories due to fatigue and muscle weakness (either in the arms/hands for feeding or in the mouth and throat for chewing/swallowing), it is the "perfect storm" for the development of **malnutrition (Figure 3). This leads to weight loss (negative caloric balance) and further muscle wasting that extends beyond the breakdown of muscles by the disease process of ALS itself** (Plowman, 2014).

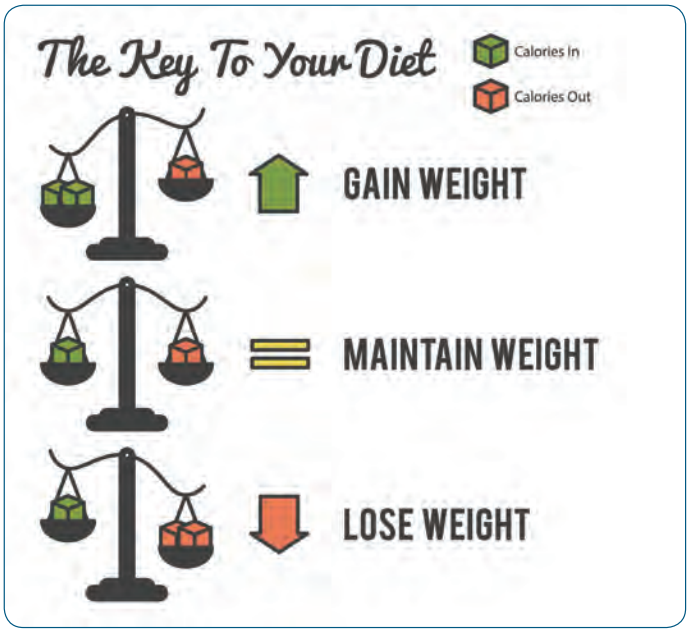

Figure 3: Maintaining weight with ALS.

Malnutrition has been noted to negatively impact disease progression and quality of life in persons with ALS (Greenwood, 2013) **and increases the risk of death by almost eight times** (Desport et al., 1999). Research also shows that difficulty swallowing by mouth creates a significant burden, longer mealtimes, and reduced enjoyment of eating, all of which contribute to weight loss (Park et al., 2013; Tabor et al., 2016).

A **feeding tube is a great option** that may be used as an *additional* source of nutritional intake and can be used while still maintaining oral intake. Placement of a feeding tube is a good way of ensuring adequate nutritional needs are met

and enhancing quality of life. Your neurologist, speech-language pathologist, and dietitian can talk with you regarding the benefits of supplemental feeding through a feeding tube. (There is more about feeding tube nutrition later in this guide.)

Takeaway Message: ***Aim to maintain your weight!***

Nutritional Interventions for ALS

Research has shown that people with ALS who maintain their weight have a longer life and better quality of life compared with those who lose weight (Miller, 2009). Talk to your doctor and dietitian about what **Body Mass Index (BMI)** should be your goal. BMI refers to the ratio of your weight to height. If excess weight is limiting mobility, gradual weight loss may be desired. Your dietitian and physician will help you meet and maintain your weight goals.

There are no specific foods or beverages that have been shown to improve outcomes in ALS. Conversely, there are none that should be universally avoided. This is where you will benefit from the skills of your ALS Center's clinical team. While the need for protein is not increased in ALS, we do not have a depot for protein, other than our lean muscle and organs. We do have storage depots for fat and carbohydrates and in order to use the protein consumed for the only job it can do (build and repair tissues, cells, etc.), carbohydrate and fat intake must be adequate or the protein will convert to energy. This is called the **protein sparing effect of dietary carbohydrates and fats**. Remember, your SLP will help you to determine appropriate food textures and consistencies, and provide strategies for your swallow. Your dietitian will be your food and nutrition coach to optimize weight, nutrition, and hydration goals. Some foods like peanut butter may not be appropriate for you to eat in a sandwich in order to avoid choking; however, you may enjoy a peanut butter chocolate milkshake with a frozen banana added. Eating certain foods and textures may be more difficult than others. Let your team know what specific foods are challenging. Together, you can choose alternatives to continue the enjoyment of your favorite foods while providing good nutrition.

Several factors increase the risk of weight loss and nutritional compromise (**Table 6**).

Table 6: Factors that Challenge Energy Balance in ALS

Increased Energy Needs	Decreased Energy Intake
Increased effort required for routine tasks such as shopping and food preparation, dressing, and eating	• Chewing • Swallowing changes • Fear of choking may limit intake
Increased difficulty in breathing as a result of decreased respiratory function (breathing takes more energy)	• Increased length of meals related to chewing and swallowing challenges • Physical challenges related to getting food and beverages to mouth • Breathing difficulties may lead to less intake
Fasciculations (visible contractions of individual muscle fibers, essentially exercise not followed by rest)	• Restricting food and fluid intake to limit toileting needs (Not recommended) • Constipation leading to less desire to eat • Depression resulting in loss of appetite

It is of utmost importance to avoid the development of malnutrition to stay "ahead of the curve" and to ensure optimal nutritional intake!

Strategies to Maintain Weight

Eat adequate calories and protein: While research does not indicate an increased need for protein in ALS, it is important to consume adequate calories or the protein that you consume is used for calories. Your body has storage depots for carbohydrate (liver and muscle glycogen) and fat (as body fat), but the only storage for protein is in your lean muscles and organs. Consuming adequate calories allows the protein that you consume to do the work of building and repairing, which is its unique function. If you do not consume adequate calories, your body will resort to breaking down energy stores in your muscles and organs. If traditional meats and protein sources are difficult to consume, other protein sources can supply the requirements. Think milk, cheese, eggs, legumes, creamy quinoa, etc.

Modify textures: Moisten foods with gravies, sauces, and dressings and eliminate crumbly dry textures, stringy consistency, etc. Choose foods that are softer, and moister.

Enhance calories: Add extra calories by using olive oil (drizzle on prepared food, even if you cook with it: 45 calories per teaspoon, with a nutty delicious flavor), coconut oil, avocado, guacamole, shakes, cream sauces, cream-based soups, cheese, or cream cheese. Consult with your physician or healthcare provider to ensure the proper type of calories is consumed.

Add shakes, smoothies, and supplements: Supplements such as Boost® and Ensure®, high calorie shakes, the addition of calorie-dense products (e.g., Benecalorie®, ice cream, protein powder) and other items that provide the most "bang for your buck" (i.e., high calories with minimal effort and energy expenditure to consume) are good choices. These suggestions should be incorporated early in the disease to reduce the chances of malnutrition from occurring (Plowman, 2014). **Ask your dietitian for recipes** and experiment with adding frozen bananas, yogurt, sweet potatoes, canned pumpkin, and peanut butter to keep it interesting and avoid boredom with the same flavors.

Reduce length of mealtimes: Extended mealtimes increase fatigue! Choose smaller, more frequent meals and snacks and use supplements as needed.

Bonus: Try taking medications in pudding, applesauce, or yogurt and one at a time, instead of a handful at once. This will provide several servings of extra calories in a day!

Here are some calorie-boosting strategies to try:

- Drizzle EVOO on foods after cooking (45 kcal/tsp)
- Add cheese, cream cheese, or whipping cream
- Add nut butters and malted milk to shakes
- Eat avocados
- Top sweet potatoes with coconut oil

Staying Hydrated

When we are dehydrated, even just 1.5% loss of body water, our mental and physical function declines. **Your dietitian can help you determine how much fluid you need.** Using certain types of cups may help to increase fluid intake.

Monitor urine concentration for adequacy of hydration, focusing on color and odor, along with bowel pattern. Urine may look concentrated and smell strong following vitamin and medications or your first-morning void.

Cups that may help with drinking:

- **Nosey Cup** has a cut out on the non-drinking side so that they can be tilted without interference by the nose. This allows the drinker to avoid tilting the head back, thereby minimizing the chance of liquid entering the respiratory tubes and causing choking.
- **Dual Cup™** is a disposable cup with a perforated cutout on the rim, providing room for the nose. This cup is ideal for people who have difficulty swallowing, as it allows one to drink without tilting the head.
- **Provale™ Cup** is available in both 5cc and 10cc delivery. The cup is designed to prevent large gulps of liquid, thus promoting safer swallowing.
- **Sip and Tip** valve cups help straws stay full of liquid, reducing the amount of air ingested and effort required. The one-way valve can be trimmed at designated areas, giving the user selective flow restriction.

Appropriate fluid intake helps with:

- Airway clearance by keeping mucous thinner (easier to cough up thin vs. thick, sticky mucus)
- Regularity of bowel movements (avoid/manage constipation: see more below)
- Kidney and urinary tract health
- Metabolism of food and medication
- Saliva/secretion management (Sometimes people with ALS are bothered by excess saliva yet the quality of the saliva does not provide comfortable moisture in the mouth. Adequate water/fluid consumption will optimize the lubricity of your saliva. Commercial rinses, toothpastes, and sprays contain enzymes present in natural saliva that may increase your oral comfort.)

...But, fluid intake may present the greatest oral challenge when there are difficulties with swallowing.

To reduce the need to drink fluid, consume these foods, which contain a high percent of water:

- Canned and fresh fruit
- Vegetables and vegetable juice
- Nectars
- Smoothies

- Cream soups
- Drinkable yogurts
- Purées in ice trays

Avoiding Constipation

Reduced physical activity may contribute to **constipation**. Mobility challenges may limit toileting independence. Some people with ALS may limit fluid intake to decrease the need for toileting assistance. But, **poor hydration can lead to constipation and discomfort, so it is important to stay hydrated.**

Fiber helps maintain regularity of bowel movements and should be included in your diet. Any discussion of fiber should include adequate fluid intake as well. To demonstrate the problem, consider mixing a psyllium-based fiber supplement with less than the recommended amount of water. Wait a few minutes; soon you will find a near concrete set up in the cup. The same scene can play out in your Gastrointestinal (GI) tract if you push fiber without enough water.

Fruits, vegetables, and whole grains are excellent sources of fiber. Your dietitian can help you select those that fit your preferences and needs. The role of fiber may change with disease progression. Some people who are less mobile may benefit from less fiber and more agents that draw water into the bowel, such as polyethylene glycol or lubiprostone to manage constipation.

Feeding Tube Option to Maintain Nutrition

Why and When to Consider a Feeding Tube

There are several parameters to consider with respect to implementing a feed tube. It is one of those situations where it probably cannot be done too soon to help with assuring proper nutrition is maintained.

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

If you are struggling to consume enough food, having trouble maintaining weight, having difficulty staying hydrated, or your breathing tests show decreased function, your ALS care team may recommend a feeding tube, either a **Percutaneous Endoscopic Gastrostomy (PEG) or a Radiologically Inserted Gastrostomy (RIG). A PEG is placed with the help of a scope that is inserted through the mouth into the stomach. A RIG is placed under the guidance of an X-ray machine. Sometimes a feeding tube is placed surgically, especially if there are changes to the anatomy or previous surgeries to the stomach or bowels.** The method of placement depends largely on practice patterns and the availability of experts at a given healthcare system. PEG placement in ALS is recommended when 10% or more body weight loss has occurred (Kasarskis et al., 1996) and/or before Forced Vital Capacity (FVC) respiratory measurements drop below 50% of the predicted value (Miller et al, 2009; Hardiman, 2000). Many centers recommend placement well before these time points.

A feeding tube offers an alternative route when feeding by mouth becomes too difficult and unsafe. Some evidence indicates that early PEG tube placement in individuals with ALS **increases survival** an average of four months (Miller et al, 1999) and in some cases, up to eight months (Spataro et al., 2011).

Having a feeding tube does not mean you can no longer eat, yet it provides a safety net. You can still enjoy the foods and beverages you want, yet use the tube to meet your fluid and nutrition goals. Your ALS doctor will refer you for the feeding-tube placement, which is generally an outpatient procedure.

The decision to undergo placement of a feeding tube is ultimately left to you and your caregivers. However, it is important to be aware of the advantages of feeding tube placement, and the benefits of early intervention. As discussed in the previous section, maintaining adequate nutrition is of paramount importance.

I didn't want a feeding tube at first, but now it is my lifeline and not so bad.

Person with ALS (Contributed by The ALS Association Alabama Chapter)

Consider your ALS care team's recommendations regarding timing of tube placement to optimize your weight and decrease the risk of the tube insertion procedure. Delaying feeding tube placement while respiratory status is declining may limit your options.

There are several videos related to decision making about the tube and its use and care. (Check out The ALS Association tube feeding video and web video http://alscare.com/feeding_tube.asp).

Feeding Tube Placement

A feeding tube that is inserted through the abdominal wall into the stomach is called a gastrostomy, or G-tube (**Figure 4**). It is held in place by either an internal balloon or bumper and an external disc or flange on the outside. The method of placement may be endoscopic (PEG), surgical, or radiologic (RIG). It is prudent to consider respiratory status and timing of tube placement as there is an increased risk with decline in breathing capacity (Miller et al., 2009).

A low-profile (skin level) gastric tube is an option for access. Similar to other gastrostomy tubes, they are held in place by either an internal balloon or bumper. These low profile tubes, often referred to as buttons, are used with extension sets of various types to allow for continuous or bolus feeding. Most providers will place a regular feeding tube first, before switching you to a low profile version after about 4 weeks.

The feeding tube is usually placed under local anesthesia where your skin is numbed with an injection. You may also be given medication by mouth or through an intravenous catheter that makes you more relaxed. Your team may ask you to **bring your Noninvasive Ventilation (NIV) device to endoscopy**, radiology, or surgery to help support your breathing during or after the procedure.

You may experience pain at the insertion site for 7-10 days. This is generally managed with over-the-counter pain medications. There should not be pain after the initial period.

Your clinic team may provide **training on how to use and care for your tube** and the insertion site. Generally, you will have home nursing to help you once you are home. **Please remember that oral care remains important** even if you are not eating by mouth.

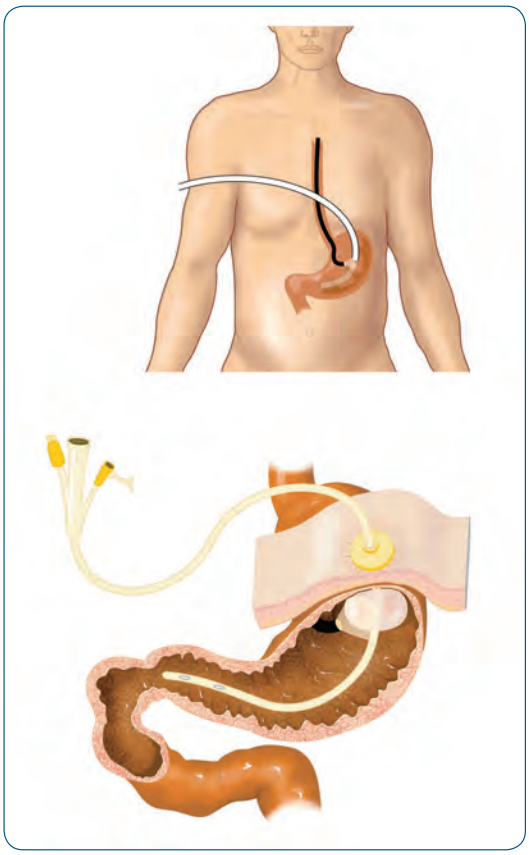

Figure 4: A regular gastric feeding tube for adults.

Feeding Delivery Methods

There are three methods by which nutrition is delivered by tube:

- **Bolus:** A defined measure of formula is placed in a syringe and flows slowly over a few minutes into the feeding tube; this is done a few times a day.
- **Gravity:** Most like natural eating patterns; nutrition is delivered several times daily over 20-30 minutes.
- **Enteral Pump:** Nutrition can be delivered at a slower rate over a longer period. Medicare and other insurers require specific situations to qualify for a pump. These medical situations usually make it necessary to administer the nutrition very slowly in order to prevent complications and/or to increase tolerability and comfort.

Feeding tubes for fluid needs as well as medication delivery: You'll want to crush non-time-released medications and/or supplements and dissolve them in warm water before flushing down the tube. Follow with ample water. Water and thin liquids may present a challenge if you have swallowing difficulty. Your dietitian can help you determine your fluid needs.

Vent the feeding tube: Certain respiratory device settings may force air past the diaphragm and into the stomach. This is called "air swallowing" (medical term: **aerophagia**) and can result in bloating and gas. You may feel full even if you have not eaten or you may experience early satiety. If you are unable to release the swallowed air by burping, the air will travel through your bowels and will be released as gas. Alternatively, you can vent the stomach directly by attaching an open syringe to your feeding tube (Martin, 2010). Follow instructions provided to vent your tube to relieve gas and bloating and increase tolerance to food and enteral (tube feeding) nutrition.

If you experience hunger pangs, you should eat or have a feeding. People tolerate portions of food or enteral nutrition differently and **your feeding method and schedule should be individualized to optimize your enjoyment and comfort**.

Some centers and hospice programs may augment your oral intake with IV hydration or nasogastric feeding for comfort.

Insurance Coverage and Tube Feeding Supplies

Medicare has very strict requirements to qualify for enteral nutrition reimbursement. If you qualify, they will cover 80%; a secondary policy may pick up the remainder. People with ALS usually qualify under the dysphagia diagnosis (difficulty swallowing).

If you have access to **VA benefits**, you can receive your enteral formula and supplies through them. Many insurance policies exclude formula from coverage as they consider formula as food and expect you to pay for your own food, although they may cover supplies. Check with your policy. Other sources of support for enteral nutrition include the Oley Foundation, and possibly nonprofit loan bank supplies and formula.

Can I put regular food in my feeding tube?

Caregiving in ALS is time intensive. Cooking and preparing food to go in the feeding tube takes effort and care to provide safe and sanitary feedings. You can feel confident that your nutritional needs can be met by commercially-prepared enteral formula. If you would like to blenderize homemade food or choose a commercially-prepared blended product, work with your dietitian to meet your goals.

SUMMARY STATEMENT

This resource guide covered information regarding the ways the normal swallowing process may become impaired by ALS and how a speech-language pathologist plays a role in assessing your swallowing in order to provide dietary modifications and safe swallowing strategies. You learned about the importance of maintaining nutrition and ways of supporting your intake, including the option of a feeding tube placement.

It is helpful to discuss these topics with loved ones, caregivers, and the medical team in order to manage your symptoms and preserve the best quality of life.

RESOURCES

Safe Swallowing for PALS: What I Need to Know and Why it Matters
www.swallowingsystemscore.org (Click on Patient Resources and this booklet will be available for download.)

Nutrition and Feeding Tube Placement in ALS: Best Practices in Clinical Decision Making
http://www.dysphagiacafe.com/2014/10/23/nutrition-and-feeding-tube-placement-for-people-with-als-best-practice-in-clinical-decision-making/

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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 and 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 and Changes in Nutritional Management of 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 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.