

Adjusting To Swallowing Changes and Nutritional Management in ALS



LIVING WITH ALS
RESOURCE GUIDE



Adjusting To Swallowing Changes and Nutritional Management in ALS

Contributors:

Lauren Tabor, MS, CCC SLP

Emily Plowman, PhD, CCC SLP

Karen Martin, MA, RDN, LD, FAND



Table of Contents

Introduction	4
Understanding Swallowing	5
Swallowing Impairment	7
How Is Swallowing Evaluated?	12
Safe Swallow Strategies	14
Dietary Modifications	15
Preparing Food for Recommended Diet Levels: Tips for Caregivers	18
Maximizing Nutrition When You Have ALS	20
Nutritional Interventions for ALS	22
Staying Hydrated	25
Feeding Tube Option to Maintain Nutrition	29
Summary Statement	33
Resources	34
Safe Swallowing for PALS: What I Need to Know and Why it Matters	34
Bibliography	35

A NOTE TO THE READER: The ALS Association has developed the *Living with ALS* resource guides for informational and educational purposes only. The information contained in these guides is not intended to replace personalized medical assessment and management of ALS. Your doctor and other qualified health care providers must be consulted before beginning any treatment.



Introduction



Swallowing and nutrition are naturally related. Your health and nutrition are affected by your ability to swallow foods and beverages. Your **Speech Language Pathologist (SLP) and Registered Dietitian/Nutritionist (RDN)** team up with you to maintain your most functional swallow and nutrition status.

This resource guide will help you understand how swallowing is affected, what you can do to maintain nutrition for energy and strength and how to keep your airway clear of liquids and food, even when the muscles involved in swallowing are affected by ALS.

This guide covers the following topics:

- Understanding the normal swallowing process
- How swallowing problems (dysphagia) are assessed and the role of the Speech Language Pathologist
- How ALS affects swallowing and makes it difficult
- Dietary changes when swallowing becomes difficult
- Avoiding malnutrition and the role of the Registered Dietitian
- Caregiver tips
- Safe swallowing strategies
- Educational resources for safe swallowing

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: **oral preparatory, oral, pharyngeal** and **esophageal**. 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 (images courtesy of www.larianmd.com):

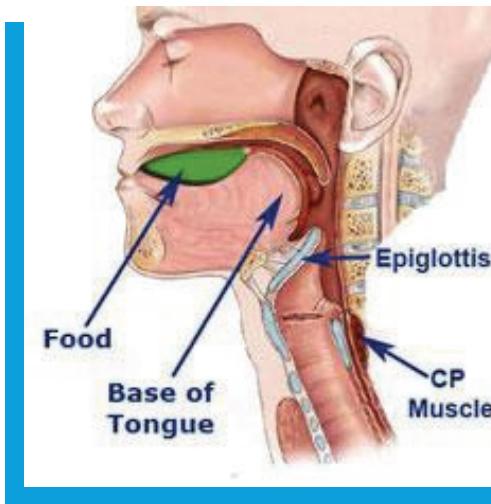


Figure 1: Oral Preparatory Stage

Oral Preparatory Stage (voluntary muscle control): 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 when 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 (voluntary muscle control): 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 particles. During the 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 toward the throat. It is important for the lips to tightly close or seal off

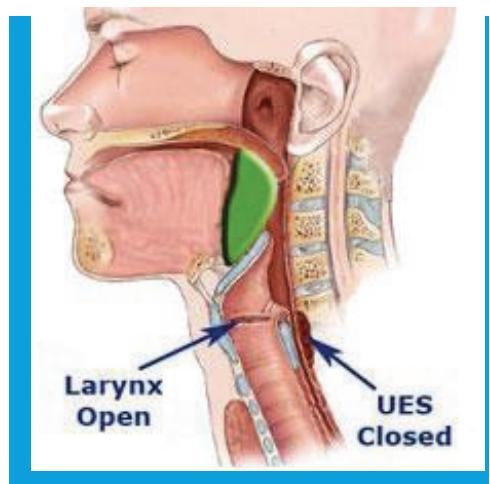


Figure 2: Oral Stage

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/liquid is directed into the throat so that it does not escape out of the mouth or the nasal cavity.

Pharyngeal Stage (involuntary muscle control):

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 the sphincter at the bottom of the throat and into the food pipe (esophagus).

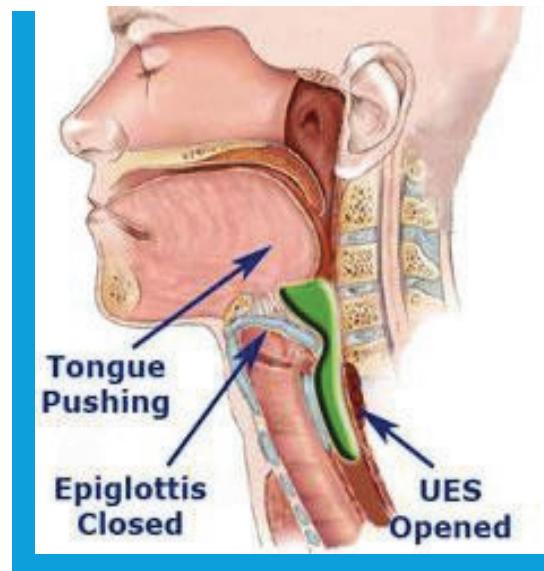


Figure 3: Pharyngeal Stage

During this stage of swallowing, the entrance to the airway (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 toward 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, including the closure of both the false and true vocal folds at the entrance to your trachea or airway.

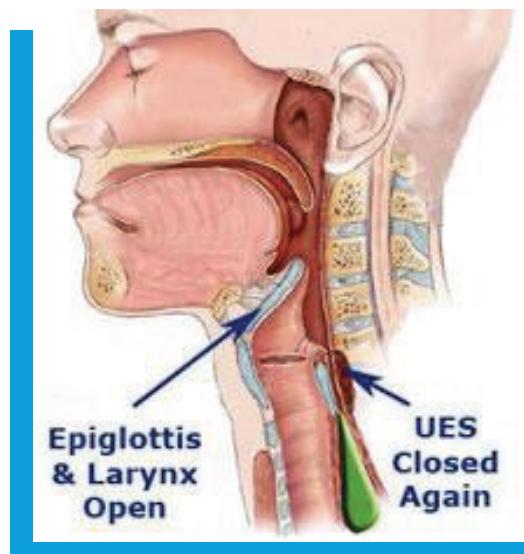


Figure 4: Esophageal Stage

Esophageal Stage (involuntary muscle control):

A sphincter (the Upper Esophageal Sphincter, UES) divides the throat from the entry to the esophagus and 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. 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 to 27 cm in length) and into the stomach.

Swallowing Impairment

Dysphagia (dis·fay·ja)

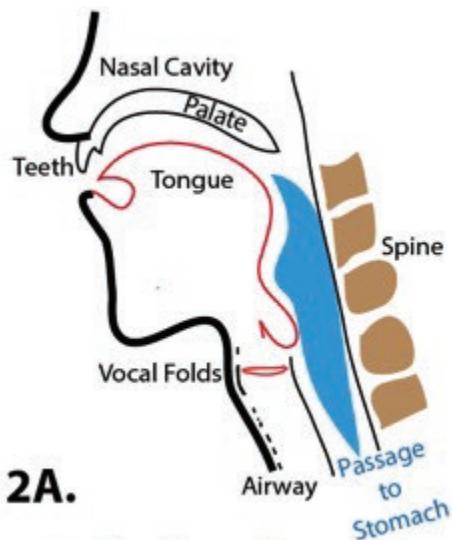
Dysphagia is the medical term for a **swallowing disorder** or swallowing impairment during any of the four stages of swallowing. Like many other medical terms, dysphagia is derived from the Greek “**dys**” meaning “**disordered**” and the root “**phag**” meaning “**to eat.**” Swallowing impairment, or dysphagia, can occur in any stage of swallowing, and often in more than one stage. Dysphagia is “an impairment of emotional, cognitive, sensory and/or motor acts involved with transferring a substance from the mouth to stomach” (Tanner, 2007). **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.” Management strategies are appropriately prescribed to target the specific type of swallowing impairments an individual is experiencing.

When treating dysphagia, it is important to evaluate the extent of the problem. That is, during what stage are swallowing difficulties experienced, or when does 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 trachea and airway. This is called **aspiration**.

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

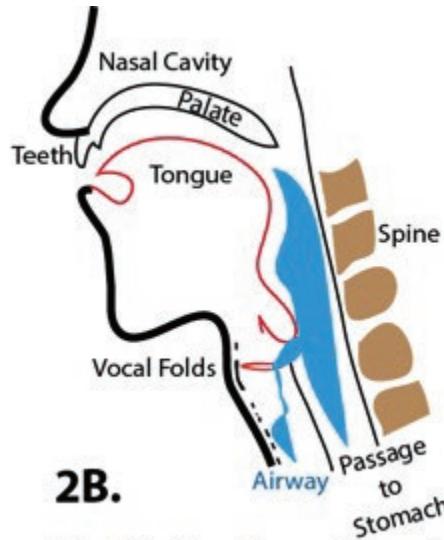


2A.

Safe Swallowing

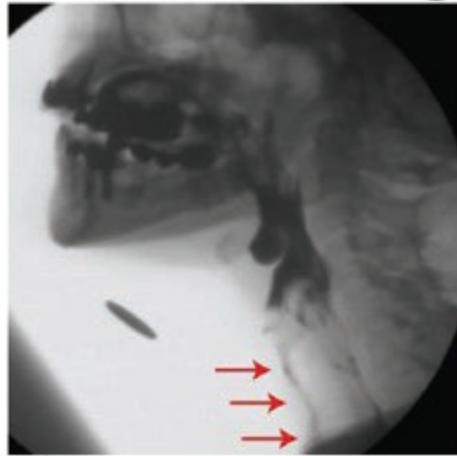


2C.



2B.

UnSafe Swallowing



2D.

Figure 5: Images of safe swallowing with liquids being routed down the food pipe with good airway protection (2A and 2C).

Figures 2B and 2D illustrate unsafe swallowing with aspiration of materials into the airway or windpipe.

Although they can co occur, it is important to distinguish a swallowing disorder from a feeding disorder to decide on the best plan for care and management. A feeding disorder involves difficulty transferring the meal from the plate to the mouth (ex: due to arm, hand or finger weakness), and can often co occur with swallowing disorders. Your occupational therapist can provide adaptive utensils, meal trays and feeding strategies to further assist with feeding related impairments.

What Causes Dysphagia?

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); ALS; multiple sclerosis (MS) and respiratory diseases.

People with ALS may experience swallowing difficulty due to the weakness and/or rigidity of the swallowing muscles and 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 expiratory pressure (breathing out) to produce a productive and effective cough.

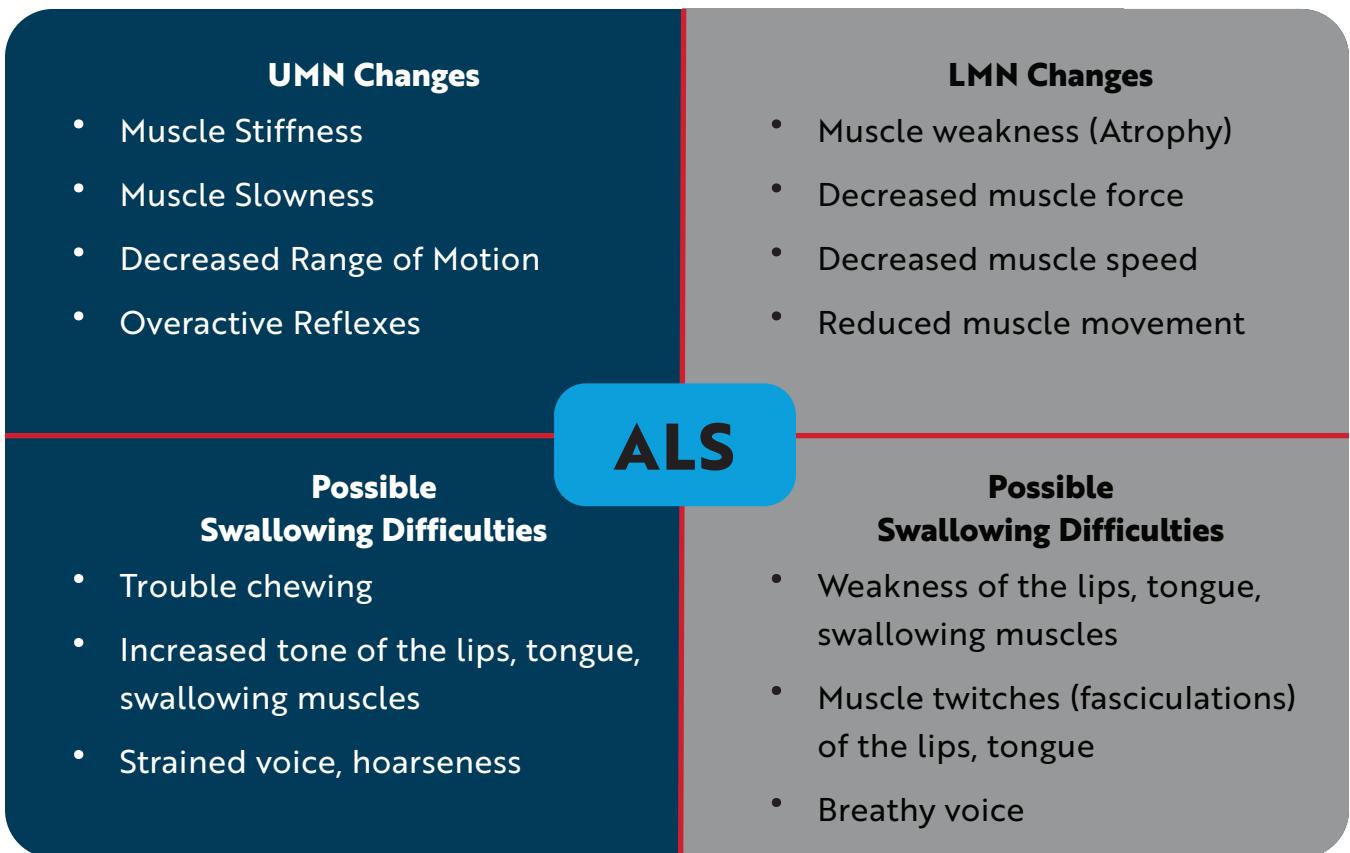


Figure 6: The symptoms of ALS divided by upper and lower motor neuron changes, with possible associated swallowing-related difficulties.

How Common Is Swallowing Impairment for People with ALS?

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

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 in Persons with 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** to keep swallowing safe.

1. During the **oral stage of swallowing**, you may experience a “**heavy tongue**” that makes it difficult to move and control food and liquid, chew and clear all material from your mouth once you swallow. Due to lip weakness, food or liquid may spill out of the front of your mouth while eating.
2. Impairments in the **pharyngeal stage of swallowing** may include a **sensation of food “sticking” in your throat** after you swallow, **coughing frequently during the meal or food or liquid coming out of your nose**.
3. Regurgitating food or liquid during or after the meal may be an indication of **esophageal related impairment**. 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** on the following page.

Table 1: Typical Swallowing Difficulties in People with ALS

Stage of Swallowing	Swallowing Difficulty
Oral Preparatory/ Oral Stage	<ul style="list-style-type: none">• 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 food/liquid to the back of the mouth• Residue in the mouth and cheek
Pharyngeal Stage	<ul style="list-style-type: none">• Food “sticking” in the throat• Food/liquid coming out of the nose• Coughing or choking during mealtime• Shortness of breath and fatigue during mealtime• Reduced cough strength and effectiveness
Esophageal Stage	<ul style="list-style-type: none">• Regurgitating food/liquid into the throat and mouth• Food “sticking” in the throat/base of neck

Risk of Aspiration and Malnutrition in Persons with ALS

Research at the University of South Florida ALS Clinic showed aspiration (material entering the airway during swallowing) occurring in approximately 30% of patients seen in their large multidisciplinary clinic. Of the patients who aspirated in their study, 58% demonstrated no cough response to food or liquid traveling into the airway (i.e., were **silent aspirators**) and 42% had a cough response that was too weak to clear the material from the airway. None of the study participants were able to effectively cough the material out of the airway. 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 detecting aspiration.

How Is Swallowing Evaluated?

The Role of the Speech Language Pathologist (SLP)

The role of the SLP is to monitor swallowing function and speech and identify changes in function throughout the disease process. Once the SLP identifies a change or decline in swallow function, they 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 finding out 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, 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 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 xray** recorded, 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. Two examples of MBS exams are provided in Figure 5: one of a patient with safe swallowing (2C) and one with unsafe swallowing (2D) under xray.

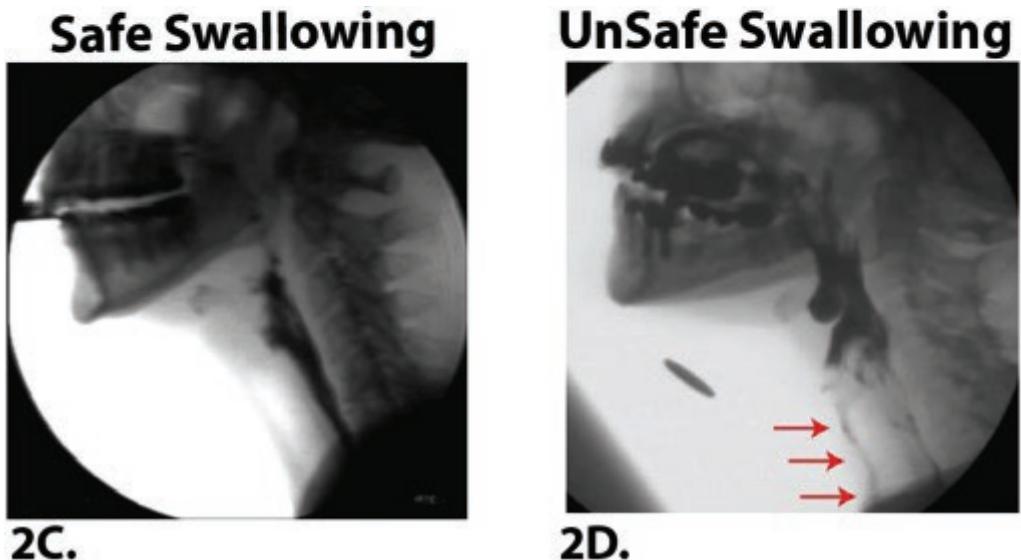


Figure 5: Image of safe swallowing with liquids being routed down the food pipe with good airway protection (2C).

Figure 2D illustrates unsafe swallowing with aspiration of materials into the airway or windpipe.

The purpose of the MBS is to assess the ability of the different muscles involved in swallowing to move the food or liquid safely and efficiently 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 patients 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 oral diet, 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 which has 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

Safe Swallowing: What the SLP May Recommend

Your SLP and registered dietitian (RD) 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 the Table 2 below:

**Table 2: Common Swallowing Strategies
Recommended by your Speech Language Pathologist**

Swallow Strategy	Description	May Help...
Effortful Swallow	<ul style="list-style-type: none">• Swallow HARD, squeezing all your throat muscles as hard as you can	<ul style="list-style-type: none">• Reduce or eliminate "leftovers" or residue in the throat
Chin Tuck Posture	<ul style="list-style-type: none">• Tucking your chin down to your chest while you swallow	<ul style="list-style-type: none">• Help protect food or liquid from entering your airway
Small Bites/ Single Sips	<ul style="list-style-type: none">• Taking small, single bites of food and sips of liquid	<ul style="list-style-type: none">• Reduce exertion and fatigue during the meal
Double Swallow	<ul style="list-style-type: none">• Swallow 2x per sip of liquid or bite of solid/soft/puree food	<ul style="list-style-type: none">• 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 of the nose
- Regurgitation
- Difficulty managing secretions (saliva) during mealtime 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 primarily 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 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 of food and sips of liquid 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 or pudding or provided in liquid form (consult your physician or pharmacist for verification). There are four different levels of modified diets reviewed in Table 3:

Table 3: Dysphagia Diet Levels and Appropriate Foods to Eat

Food Diet Level	Examples of food in this level:	
Level 1: Pureed	<ul style="list-style-type: none"> • Pudding • Pureed oatmeal, breads, meats 	<ul style="list-style-type: none"> • Hummus • Pureed fruits and vegetables
Level 2: Mechanical Soft	<ul style="list-style-type: none"> • Scrambled eggs • Meatloaf • Well-cooked vegetables 	<ul style="list-style-type: none"> • Pancakes • Mashed potatoes • Canned/cooked soft fruits
Level 3: Advanced	<ul style="list-style-type: none"> • Bread slices • Muffins • Moistened cereals • Pasta, casseroles 	<ul style="list-style-type: none"> • Baked potatoes • Soft/ripe fruits • Fish
Level 4: Regular	<ul style="list-style-type: none"> • No Food Avoidances or Restrictions • Help to eliminate food/liquid left over after the initial swallow 	

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 more viscous (thick) and slow down the flow rate of the liquid material during swallowing. Recall that swallowing occurs in less than two seconds. During this time over 26 pairs of muscles and five different cranial nerves need to coordinate and move in a complex pattern to protect the airway and direct the ingested materials toward 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.

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.

Coughing or throat clearing while drinking liquids is a sign of **aspiration**.



Table 4: Thickening Your Liquids

Type of Liquid	Description
Nectar Thickened Liquids	<ul style="list-style-type: none">Liquid is a consistency slightly thicker than waterDoes not contain fruit nectar or nectar flavoring
Honey Thickened Liquids	<ul style="list-style-type: none">Liquid resembles the consistency of honey at room temperature

Examples of thickening agents: Nestle® Resource Thicken Up, Simply Thick, Thick-It

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

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		<ul style="list-style-type: none">• Take smaller, more frequent meals throughout the day• Add snacks
Difficulty Chewing		<ul style="list-style-type: none">• Moisten foods with gravies and/or sauces• Use smaller bite size• Change food consistency to a softer diet that requires less chewing
Weight Loss		<ul style="list-style-type: none">• Add foods high in calories (e.g., peanut butter, butter, condiments)• Drink full cream milk smoothies or shakes• Supplement nutrition with Boost or Ensure drinks, Resource Benecalorie• Add snacks throughout the day
Loss of Enjoyment		<ul style="list-style-type: none">• Increase taste, temperature and textures using spices/sauces
Fatigue/Shortness of Breath		<ul style="list-style-type: none">• 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. (Adapted from Yorkston, Miller and Strand, 2003.)

To further combat the challenge of fatigue that comes with muscle weakness, work with your team's occupational therapist (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:

- The presence of **hypermetabolism** (when you burn calories faster than "normal") (Desport, 2001)
- **Reduced caloric intake** because of eating less due to swallowing problems and fatigue (Ngo, 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. This leads to weight loss (negative caloric balance) and further muscle wasting that extend beyond the catabolic nature of the ALS disease itself** (Plowman, 2014).

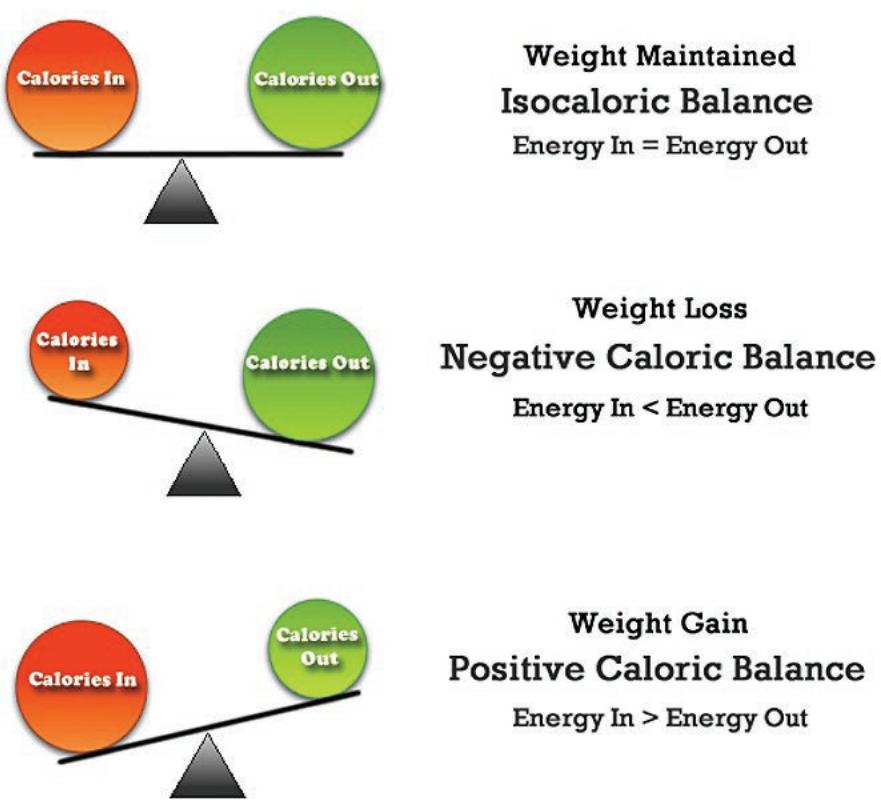


Figure 7: Maintaining weight with ALS (Image: www.cod.edu)

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** (Couratier, et al., 1999). Research also shows that difficulty swallowing by mouth creates significant burden, longer mealtimes and reduced enjoyment of eating, all of which contribute to weight loss (Park et al, 2013; Tabor 2015).

A feeding tube is another 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 has been shown to increase survival in PALS by four to six months (Spataro et al., 2011) and is a good way of ensuring adequate nutritional needs are met. Your Neurologist, Speech-Language Pathologist and Dietician can talk with you regarding the benefits of supplemental feeding through a feeding tube. (More about feeding tube nutrition later in this guide.)

Nutritional Interventions for ALS

The takeaway message is:
Aim to maintain your weight!

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. 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, but 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 enjoyment of your favorite foods while providing good nutrition.

Several factors increase risk of weight loss and nutritional compromise:

**Table 6: Reasons for Increased Energy
Needs and Decreased Intake**

Increased Energy Needs	Decreased Energy Intake
Longer Mealtime Duration	<ul style="list-style-type: none">• Chewing, swallowing changes• Fear of choking
Work of breathing related to decreased respiratory function (as % predicted FVC ↓ , work of breathing ↑)	<ul style="list-style-type: none">• Increased length of meals related to chewing, swallowing, physical challenges related to getting food and beverages to mouth and breathing difficulties
Fasciculations (visible contractions of individual muscle fibers, essentially exercise not followed by rest)	<ul style="list-style-type: none">• Restricting food and fluid intake to limit toileting needs (not recommended)• Constipation
	<ul style="list-style-type: none">• Depression

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 protein: While research does not indicate an increased need for protein in ALS, it is important to consume adequate calories or that the protein 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, rather than breaking down 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.).

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

Caloric enhancements: Add extra calories with 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 and cream cheese.

Shakes, smoothies and supplements: Supplements such as Boost® and Ensure®, high calorie shakes, the addition of calorically dense products (i.e., 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)

Calorie Boosters

- Drizzle EVOO on foods after cooking—45 kcal/tsp
- Add cheese, cream cheese, whipping cream
- Add nut butters & malted milk to shakes
- Avocado & Guacamole
- Coconut oil is great on sweet potatoes



Figure 8: Foods high in calories to help maintain weight

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 or peanut butter to avoid boredom with the same flavors.

Reduce 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!

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.**

Monitor urine concentration for adequacy of hydration, along with bowel patterns. Urine may look concentrated and smell strong following vitamins and medications or during your first morning void.

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.

Cups That May Help With Drinking:



Nosey
Cup



Dual Cup™

Provale ® 5 and 10cc



Figures 9 through 11: Cups that may aid in drinking

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 foods that contain a high percent of water:

- Canned and fresh fruits
- Vegetables
- Nectars
- Smoothies
- Cream soups
- Vegetable juices (e.g., V8)
- Drinkable yogurts
- Purees in ice trays

Don't restrict fluids!



- Condom Catheters—energy savings, travel savers
- Urinals
- Conversion Bidet

Figure 12: Devices that may aid in urination.

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 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 fluid intake as well. Consider mixing a psyllium based fiber supplement with less than the recommended amount of water. Wait a few minutes and you return to find a near concrete set up in the cup. The same scene can play out in your gastrointestinal tract (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 with your preferences and needs. The role of fiber may change with disease progression, as those that 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

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)**. PEG-tubes tend to be most common. Specifically, PEG placement in ALS is recommended when 10% or more body weight loss has occurred (Berryman et al., 1996) and/or before forced vital capacity (FVC) respiratory ratings drop below 50% of the predicted value (Miller et al, 2009; Hardiman, 2000). Many centers recommend placement well before these time points.

The PEG offers an alternative route when feeding by mouth becomes too difficult and unsafe. 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 8 months (Spataro, 2011).

Having a feeding tube does not mean you can no longer eat. It provides a safety net. You can still enjoy foods and beverages you want while using 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 the person living with ALS and their caregivers. However, it is important to be aware of the advantages of PEG tube placement and the benefits of early intervention. As discussed in the previous section, maintaining adequate nutrition is of paramount importance.

Consider your ALS care team's recommendations regarding timing of tube placement to optimize your weight and respiratory status and decrease 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 gastrostomy tube (G tube) is inserted through the abdominal wall into the stomach. It is held in place by either an internal balloon or bumper and an external disc or flange on the outside. The original placement may be endoscopic (PEG), surgical or radiologic (RIG). Evidence based practice recommendations support enteral (tube) nutrition via PEG to stabilize weight (Miller et al., 2009). It is prudent to consider respiratory status and timing of tube placement, as there is increased risk with decline in breathing capacity related to sedation and tolerance (Miller et al., 2009).

A low profile (skin level) gastric device is an option for feeding access. Like gastrostomy tubes, they are held in place by either an internal balloon or bumper. These devices, often referred to as buttons, are used with extension sets of various types: Bolus sets are used for medication administration and feeding by gravity and bolus methods. Right angle connectors are typically used for pump delivered feedings.

Feeding tubes can be utilized to meet your fluid needs as well as deliver medications. Crush non-time-release meds and dissolve in warm before

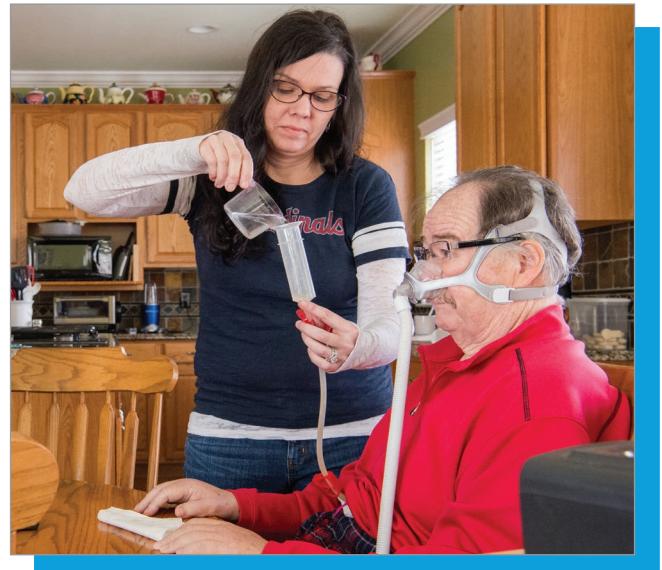
flushing down the tube. Follow with ample water. Water and thin liquids may present a challenge if you have swallowing difficulty. Your dietitian can help you determine your fluid needs.

Your team may ask you to **bring your non invasive ventilation (NIV) to endoscopy**, radiology or surgery.

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

Oral care remains important even if you are not eating by mouth.

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.



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 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 to 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. (Rate < 100 ml/hr or J tube; aspiration risk/pneumonia, gastroesophageal reflux disease (GERD), vomiting, diarrhea, dumping syndrome, circulatory overload, blood glucose fluctuations).

Vent the feeding tube: Certain respiratory device settings may force air past the diaphragm and into the stomach. This is called **aerophagia** and can result in bloating and gas. You may feel full even if you have not eaten or experience early satiety. If you are unable to release the swallowed air by burping, the air will travel through your bowels and be released as gas. Alternately, 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 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 (tube feeding) reimbursement. If you qualify, they will cover 80%; a secondary policy may pick up the remainder. People with ALS most usually qualify under the dysphagia diagnosis (difficulty swallowing). Even people with a great deal of weight loss from other issues may not meet Medicare guidelines depending on the reason for the weight loss.

If you have access to **VA benefits**, you can receive your enteral formula and supplies through them; though, they do not currently supply extension tubes for low profile gastric devices.

Many insurance policies exclude formula from coverage because 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: Loan bank, Oley Foundation, etc.

Summary Statement

We discussed a lot of information regarding:

- The swallowing process
- Normal and impaired swallowing
- How ALS affects swallowing
- Dietary modifications and safe swallowing strategies
- Importance of maintaining nutrition
- The option for feeding tube placement

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

The goal in writing this resource guide is to provide you with the knowledge and awareness you need to advocate for yourself and ensure your symptoms are appropriately managed. Please take time to read the resources provided in the links below. Should you have questions regarding the information discussed in this manual, please contact your local SLP or RD.

Resources

Safe Swallowing for PALS: What I Need to Know and Why it Matters

https://www.neals.org/uploads/blog/doc/Safe_Swallowing_for_PALS.pdf

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/>

Bibliography

- Carpenter, R.J., 3rd, T.J. McDonald, and F.M. Howard, Jr. (1978). The otolaryngologic presentation of amyotrophic lateral sclerosis. *Otolaryngology*, 86(3 Pt 1): p. ORL479 to 484.
- Chen, A. and C.G. Garrett (2005). Otolaryngologic presentations of amyotrophic lateral sclerosis. *Otolaryngol Head Neck Surg*, 132(3): p. 500 to 504.
- Desport, J.C., et al. (2001). Factors correlated with hypermetabolism in patients with amyotrophic lateral sclerosis. *Am J Clin Nutr*, 74(3): p. 328 to 334.
- Desport, J.C., et al. (2005). Hypermetabolism in ALS: correlations with clinical and paraclinical parameters. *Neurodegenerative Disease*, 2(3-4): 202 to 207.
- Greenwood, D. I. (2013). Nutrition Management of Amyotrophic Lateral Sclerosis. *Nutrition In Clinical Practice*, 28(3), 392. doi:10.1177/0884533613476554Dis, 2005. 2(3-4): p. 202 to 207.
- Groher, M and Crary, M. *Dysphagia: Clinical Management in Adults and Children*. Maryland Heights, MI: Mosby Elsevier.
- Kasarskis, E. J., and Berryman, S. (1996). Nutritional status of patients with amyotrophic lateral sclerosis: Relation to the proximity of Death. *American Journal of Clinical Nutrition*, 63(1), 130.
- Martin, K., Jackson, L. (2010). Management of Enteral Feedings for Respiratory Compromised Patients on Noninvasive Ventilation. *Nutrition in Clinical Practice*, 25 (1), Abstract 34.
- Miller, R.G., et al. (2009). *Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence based review): report of the Quality Standards Subcommittee of the American Academy of Neurology*. Neurology, 73(15): p. 1218 to 1226.

Mitsumoto, H. (2009). ***Amyotrophic Lateral Sclerosis: A Guide for Patients and Families.*** New York: Demos Health.

Ngo, S.T., F.J. Steyn, and P.A. McCombe (2014). Body mass index and dietary intervention: implications for prognosis of amyotrophic lateral sclerosis. ***J Neurol Sci***, 340(1-2): p. 5 to 12.

Paris, G., Martinaud, O., Petit, A., Cuvelier, A., Hannequin, D., Roppeneck, P., and Verin, E. (2013). Oropharyngeal dysphagia in amyotrophic lateral sclerosis alters quality of life. ***Journal of Oral Rehabilitation***, 40(3), 199. doi:10.1111/joor.12019.

Plowman, EK (2014). Nutrition and Feeding Tube Placement for PALS: Best Practice in Clinical Decision Making. ***Dysphagia Café***, from <http://www.dysphagiacafe.com/2014/10/23/nutrition-and-feeding-tube-placement-for-people-with-als-best-practice-in-clinical-decision-making/>.

Rosen AD (1978). Amyotrophic lateral sclerosis: clinical features and prognosis. ***Ann Neurol*** 35: 639 to 642.

Spataro, R. et al. (2011). Percutaneous endoscopic gastrostomy in amyotrophic lateral sclerosis: effect on survival. ***J Neurol Sci***, 304(1-2): p. 44 to 8.

Yorkston, K., Miller, R., and Strand E. ***Management of Speech and Swallowing in Degenerative Disease, 2nd edition.*** Austin, TX: Pro-ed, Inc.

Notes

Notes

Notes



The ALS Association
1300 Wilson Boulevard
Suite 600
Arlington, VA 22209
[www\(ALS.org](http://www(ALS.org)

About The ALS Association

The ALS Association is the largest philanthropic funder of ALS research in the world. The Association funds global research collaborations, assists people with ALS and their families through its nationwide network of care and certified clinical care centers, and advocates for better public policies for people with ALS. The ALS Association is working to make ALS a livable disease while urgently searching for new treatments and a cure.