A Quick Reference Guide to ALS Care for Home Health, Hospice, and Skilled Nursing Facilities

Respiratory

The majority of ALS patients will eventually experience difficulty breathing. Unlike typical patients with respiratory problems, ALS patients rarely have trouble pulling oxygen out of the air they breathe and therefore the use of supplemental oxygen is typically not appropriate. The problem ALS patients face is simply the inability to move a sufficient amount of air in and out of the lungs due to respiratory muscle weakness. While this can result in low blood oxygen levels, it also results in an equally dangerous buildup of carbon dioxide in the lungs and blood, which the use of supplemental oxygen does not address. The solution to respiratory problems in ALS patients is either non-invasive mechanical ventilation called NIPPV or BIPAP, or invasive ventilation called a ventilator.

BIPAP

- Bipap- bilevel positive airway pressure- is the name brand of a non-invasive positive pressure ventilator (NIPPV or NIV).
- Used with a mask that must be snuggly strapped to the patient's head. There are several different types of masks, some over the mouth and nose and some just over the nose.
- Positive pressure means air is pushed in; bilevel means there are different pressures for inhaling and exhaling, referred to as IPAP and EPAP.
- Some bipap units, labeled "ST", can have a rate set on them so that the patient gets a set number of breaths.

AVAPS

- Avaps-average volume pressure assured support.
- Similar to the bipap ST, but also has a volume setting so the air that it provides is more consistent with the amount an individual needs (based on weight).
- Found to be more effective and better tolerated by ALS patients.

TRILOGY

- Trilogy is a mechanical ventilator that provides both volume and pressure ventilation and delivers the therapy either invasively (via a trach) or noninvasively (via a mask).
- Advantages over bipap or avaps:
  - Can be more finely tuned to patient's needs
  - Has an internal battery that can last up to 6 hours. Bipap and avaps do not have batteries- they must always be plugged into an outlet or with an adapter into the cigarette lighter in a car or hooked to a portable battery that weighs about 40 pounds.
- Disadvantages over bipap and avaps:
  - More costly. Some insurance will not approve for non-invasive use unless the patient is nearly 100% dependent on it (20-24 hours per day).
  - More costly for hospice to pay for.

Summary

All of these devices, when used non-invasively (with a mask), have limited ability to assist with breathing. As the respiratory muscles weaken due to ALS, even these devices cannot sustain life. They are all appropriate for hospice ALS patients, but cost may be the limiting factor for hospice particularly with the Trilogy.

Oxygen is NOT for Hypoventilation in Neuromuscular Disease

By E.A. Oppenheimer, MD, FCCP

If progressive respiratory failure occurs in people with neuromuscular disease, an abnormal nocturnal oximetry study is often an early indication that hypoventilation is occurring. There are significant periods of decreased oxygen levels in the blood or hypoxemia during sleep when lying flat, in addition to decreases in vital capacity (VC), maximum inspiratory force (MIF) and maximum expiratory force (MEF). Decreased oxygen saturation (SaO_2) combined with increasing carbon dioxide (CO_2) retention or hypercapnia are the hallmarks of hypoventilation. This is sometimes called ventilatory pump failure, due to the weakened respiratory muscles.

Patients with neuromuscular diseases who are developing progressive respiratory failure due to respiratory muscle weakness will die unless mechanical ventilation is used. The rate of progression is often hard to predict. Some patients seem suddenly to experience life-threatening hypercapnic respiratory failure. They may not have been aware of gradually increasing symptoms and signs, particularly since they are often not physically active and are often not being regularly monitored with simple pulmonary function tests.

Administering oxygen does not provide assistance to the weakening respiratory muscles, but gives both the patient and the doctor the false impression that appropriate treatment is being provided. While in fact hypoventilation is mistaken for an oxygen transfer problem. Indeed, administering oxygen can mask the problem. Also there is a danger of causing respiratory depression by giving oxygen. Oxygen is not the treatment for hypoventilation. It will improve the SaO_2, but not the hypoventilation and may increase the danger of dying of sudden respiratory failure.

In hypercapnic respiratory failure due to hypoventilation, the SaO_2 falls due to the rise of the CO_2. The alveoli in the lungs (tiny gas exchange units) should clear most of the CO_2 out with each breath. Instead, with hypoventilation, CO_2 accumulates and thus there is decreased room in the alveoli for oxygen. When mechanical ventilation using room air is provided, it lowers the CO_2 in the alveoli, corrects the SaO_2 and rests the respiratory muscles. The ventilator should be adjusted to achieve a normal SaO_2, on room air. If oxygen is being administered, one cannot use noninvasive oximetry to tell whether enough assisted ventilation is being provided; repeated arterial blood gas specimens (ABGs) would be needed.

When there is respiratory failure in neuromuscular patients (ALS, postpolio, SMA, muscular dystrophy, etc.) who have no additional pulmonary disease that impairs oxygen transfer, the ventilator set-up is adjusted to:

- be comfortable for the patient;
- achieve SaO_2 of 95% or higher on room air (this can be measured with a finger-sensor oximeter);
- Assist the patient to effectively cough and clear secretions;
- provide improved oral communication (if vocal communication is possible).

It has been common for people using noninvasive nasal ventilation (NPPV) with a bi-level positive pressure unit to use inadequate settings; frequently, they are not monitored with clinical evaluation and oximetry. The EPAP is often set too high - usually it should not be higher than 3-4 cm H_2O; the IPAP is set too low - usually it needs to be 12-16 cm H_2O and adjusted to achieve an oxygen saturation of 95% or higher. Some situations may require administering oxygen, such as pneumonia due to infection or aspiration. If this occurs in patients with respiratory muscle weakness and hypoventilation, then it is important to provide both assisted ventilation and supplemental oxygen, and use ABGs to monitor them.

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This article was reprinted with permission from Edward Anthony Oppenheimer, MD, FCCP. It was originally published in the International Ventilator Users Network IVUN News, Spring 2000, Volume 14, Dr. Oppenheimer is Associate Clinical Professor of Medicine, University of California, Los Angeles (uCLA). He retired after 31 years with Kaiser Permanente Medical Group where he organized and coordinated the Kaiser home ventilator program. Although Dr. Oppenheimer is no longer seeing people in office practice and does not have an office referral, he will try to respond to comments and questions emailed to him at Eaopp@UCLA.edu.

Secretion Management in ALS

Some patients with ALS experience problems with **excessive oral secretions** or experience **thick phlegm that they cannot cough up**. Excessive secretions are experienced when the weakened muscles of the mouth, tongue and throat are not able to swallow the normal oral secretions. Secretions may pool in the mouth and cause drooling, or may collect in the back of the throat where they may cause choking or aspiration into the lungs, increasing the risk of pneumonia. Controlling the secretions is important to decrease the chance of respiratory complications.

Drying Secretions

- Medications commonly prescribed to decrease saliva production include amitriptyline (Elavil®) and other tricyclic antidepressants, glycopyrrolate (Robinul®), and scopolamine (Transderm Scop®) patches.
- Possible side effects of these medications include dry mouth, constipation, and urinary hesitancy. Less common side effects are sedation and confusion.
- Atropine drop in the mouth may be helpful in some cases.
- Botulinum toxin (Botox®) is often prescribed if the medications noted above are ineffective. Botulinum toxin is injected by a physician directly into the parotid and submandibular glands, where the saliva is made, causing a decrease in saliva production. It often takes 1-2 weeks for maximum effectiveness, and if it is successful, the effects usually last up to 12 weeks.
- For sleeping difficulties due to the build-up of saliva, try elevating the head of the bed with pillows or by placing a bed wedge under the head.

Excessive nasal secretions

- Drip and cause build up in the back of the throat.
- Try over the counter anti-histamines such as Claritin® or Allegra®.
- If these don't work, then we advise that you contact your primary care physician for a prescription form of an antihistamine.
- There are some that are available in liquid form, such as levocetirizine (Xyzal®) which may be effective for nasal congestion.

Thinning Secretions

- Thick secretions are caused by several factors 1) drying of secretions with medications; 2) low airflow as a person's breathing capacity declines 3) loss of diaphragm muscle strength which diminishes the ability to cough. Management of these "thick secretions caught in the back of the throat" often requires several approaches in combination:
- Make sure you are adequately hydrated
- Guaifenesin (which is contained in Robitussin Chest Congestion® or Mucinex®) can be taken orally or (if in liquid form) given through a feeding tube. Robitussin is taken as 1-2 teaspoons every 4 hours. Mucinex is a 600 mg pill, taken twice a day. Guaifenesin dose is 400mg every four hours. If prescribed as liquid, this usually is 4 teaspoons every 4 hours.

- Other medications that break up thick secretions are Albuterol, Mucomyst or DuoNeb. These nebulized treatments can be taken up to four times a day and should be performed on a routine basis to prevent build up of secretions in the back of the throat or lungs.
- A suction machine is used to remove secretions

Mobilizing Secretions

- Perform an augmented manual cough with a bag or with abdominal pressure.
- Following a nebulizer treatment, a mechanical insufflator-exsufflator (CoughAssist®) device can help the person cough up the secretions. The insufflator-exsufflator machine produces a pressure that helps the person with weakened muscles produce a stronger, more effective cough.

- Problems with thick secretions after combining all these approaches a high-frequency chest wall oscillation (VEST®) device may be considered.

This is a rapidly oscillating device that resembles a vest, and dislodges mucus from the bronchial walls and mobilizes secretions and mucus from the smaller to larger airways where it can be cleared by coughing or suctioning

Oral Care for the ALS Patient

Assistive devices

Toothpaste dispenser-- Eliminates the need to squeeze paste from a tube. For patients with decreased hand function, a tube or pump of toothpaste is inserted into the holder and paste is placed on the toothbrush by depressing a large handle on the dispenser (available through catalogs).

Floss holders-- Basic designs available at stores. The "Y" shaped plastic holder includes a handle, a knob to secure floss, and two grooved extensions to guide floss into a short horizontal position. The handle is held while the horizontal section of floss is slid between the teeth.

Velcro or metal hand and wrist cuffs-- Can be fitted around the hand or wrist. Toothbrush attaches to cuff and can be moved with arm action. Some cuffs have a self-rotating brush attached to make maneuvering the toothbrush easier.

Electric toothbrushes-- Eliminate need for extensive arm action. Many models are designed with large handles to make gripping easier. Disadvantages may be that the device is too heavy to lift; handles can become slippery and difficult to grip. But these brushes may make plaque removal easier, and their use is not discouraged.

Handle build-ups-- A very inexpensive aid that makes gripping handles easier. No purchase is necessary. 1. A racquetball or tennis ball can fit over the end of a brush handle after one cross cut has been made into the ball. 2. Styrofoam tubing (available through catalogs from healthcare providers) can fit over skinny handles of regular toothbrushes.

Extended handles-- Provided through catalogs; ask an occupational therapist for availability. Long metal or plastic rods that hold the toothbrush at mouth level while the arm, remains at waist or chest level, provides the movement.

Proxabrush-- For use once self-care is too difficult. This toothbrush-like handle holds a small cylindrical- or cone-shaped brush and is best for cleaning open spaces between teeth, around isolated teeth and under fixed appliances such as bridges (available in stores).

Rinses

- **Oral-B anticavity-alcohol free** (which is over the counter),
- **Peridex or Periguard**-Prescription strength **Chlorhexadine rinse** (you need an order from your dentist). This helps to treat gingivitis.

Plak-vac toothbrush- suction toothbrush

- Attached to a suction machine
- This device allows water and toothpaste to be removed as the teeth are being brushed.

Assistance with Brushing /Flossing

- Certain positions will make this task easier. While in your wheelchair have your caregiver stand in front of you or to the side and face them. If there is not a headrest, you may want to have the chair backed up to the wall to prevent your head from falling backward. They should use one hand to stabilize the chin and pull open the lips and the other hand to brush with.
- When flossing, there are two positions that should be considered as well. First, your caregiver should stand behind your wheelchair and wrap their arms around your head and lean over looking into your mouth. The second choice is to floss while you are lying in bed with your head up and stand a little behind and to the side of your head. This position decreases the strain on your neck. A floss holder may be useful if regular flossing becomes too difficult.

Difficulty and/or slurred speech

(From ALSA-Living with ALS: Adjusting to swallowing and speaking difficulties)

Slurred speech is a symptom of **dysarthria**, a neurologically-based speech disorder that results in weakness or spasticity of the lips, tongue, jaw movement, soft palate, and respiratory muscles.

Because ALS is a progressive disorder, the slurred speech may eventually become severe, particularly if it is present early in the disease.

Bulbar symptoms

- Weakness and lack of coordination of the muscles that control speech, swallowing, the ability to maintain an open upper airway, and the ability to clear away saliva.
- Involve the part of the brain known as the **medulla oblongata (the "bulb" like structure)** at the top of the spinal column.

In order to adjust to these weakened muscles in the mouth, you can make changes in how, where and when you speak and what you do to be understood. In general, it will be easier for people to understand you:

- While facing your listener in a place that is quiet and well lit. This arrangement helps listeners hear you more clearly, as well as see your mouth and facial expressions.
- Recognize that your speech may be clear in one situation but not in others, for example when you are tired, emotionally stressed, or when there is background noise.
- Try getting your listener's attention before you start; say the person's name or give a tap on the shoulder, so he/she is ready to pay close attention.
- You also should be prepared to shift to writing as needed.
- State your topic first in order to help listeners understand the meaning of what you are saying, even if they do not catch every word. Sometimes your speech may be more easily understood if you write the topic or key words, such a "medicine."
- Ask your listeners if they understand you; sometimes people will pretend to do so because they do not want to embarrass you or themselves. If you are unsure, ask them to repeat or explain what you said. This effort, by the speaker, generally is a very welcome suggestion for the listener.

Tips for Speaking Difficulties

The following are some very specific tips that can help you with your speaking difficulties:

- Speak slowly and carefully; repeat your words if necessary.
- Convey your message in as few words as possible, realizing that those at the end of a sentence are lost more easily.
- Carefully pronounce all the syllables in words; if you have trouble speaking slowly, tap out each syllable with your finger as you say it, for example, "re-frig-er-a-tor."
- Emphasize the final sounds of each word, since slurred speech can omit them, for example, boo"k" or ha"t."
- Take a breath before each phrase or set of words, because breath is the power behind your voice, making your words easier to say and hear.
- Say your most important words more loudly by taking a breath first.

Helpful Hands, Helpful Voices

Your family and friends can help you compensate for your speaking difficulty.

- They can explain to others that you may need additional time to respond, but that you understand everything and should be included in conversations.
- Moreover, your friends can emphasize to your listeners the importance of being straightforward and sincere.
- Because your regular speaking partners are more familiar with your speech, they may be able to translate for you.
- In addition, arrange for someone to maintain and transport any assistive communication devices that your might have.
- A communication partner who provides moral support and serves as your advocate is extremely valuable.

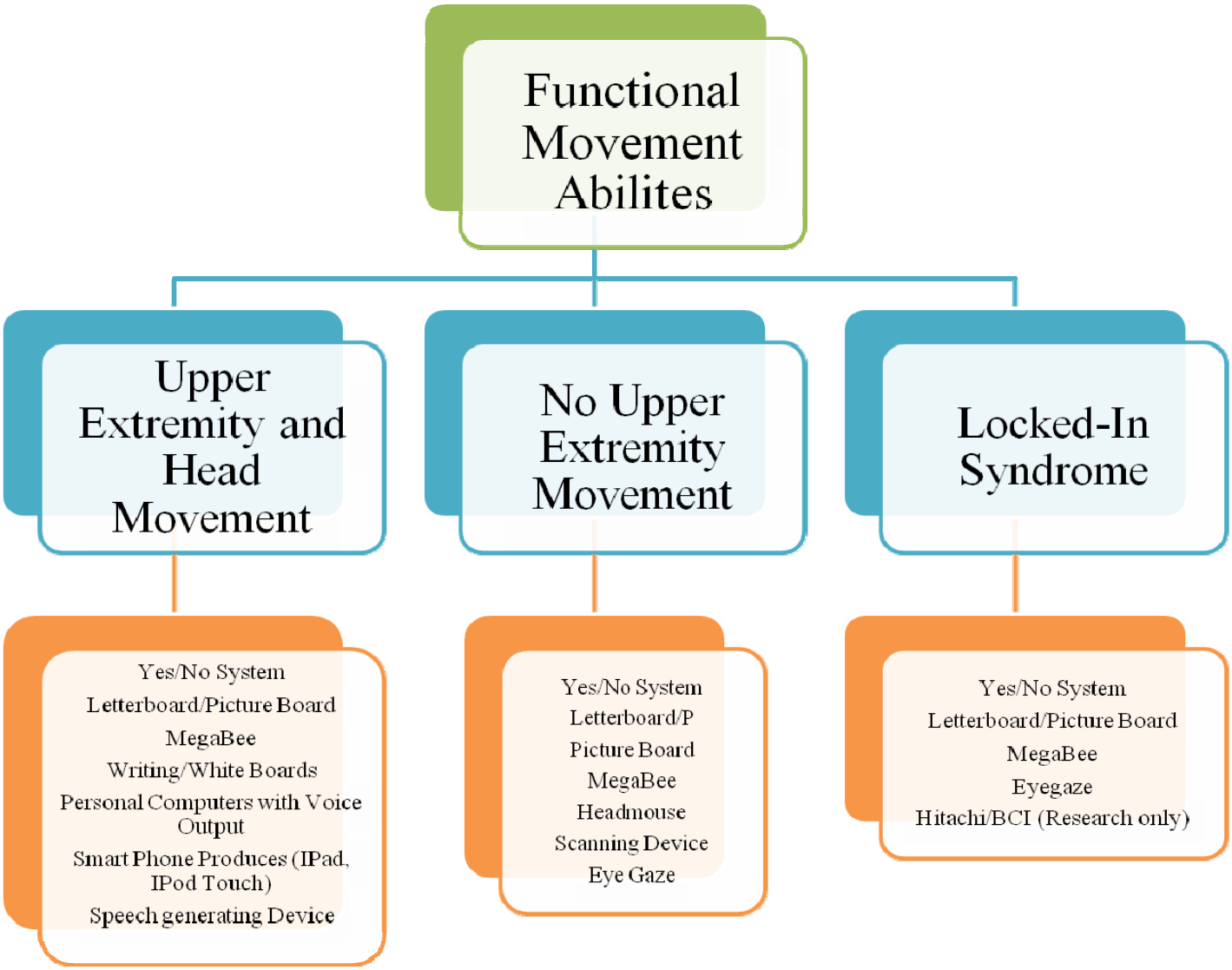
To be an effective speaking partner for a person with ALS:

- Check your hearing. If you have hearing loss, recognize that it will be a bit more challenging to understand slurred speech, so please use a hearing aid or amplifier.
- Give the person your full attention and concentrate on his/her face before you start listening. It is easier to understand the person when it is quiet and you can see their face.
- If you do not understand something, ask the person to please say it again slower-or-louder.
- Have the speaker spell words that are not clear.
- Remind him/her to state when the topic is changing so you will know the context.
- Give feedback about what you did hear correctly, so that the person can fill in the missing word(s); for example, "You want to go for a drive, but I missed where."
- Use an **alphabet board** (described later) or write the alphabet on a 5"x7" index card; point to the first letter of each word you say.
- If you have repeated the words twice and they still are not clear, spell them out loud.

Using the telephone

With ALS, talking on the phone may be challenging because you cannot use your face and gestures to assist your speech. Here are some practical suggestions to improve your ability to be understood on the phone:

- Right away, tell the listener that you have a speech disorder and that you will repeat a word or phrase if necessary.
- A **weak-voice amplifier** can increase the loudness of your voice over the phone.
- A **speaker phone** allows you to talk without using a hand held receiver; it enables you to be part of any conversation because a nearby friend can clarify or repeat what you said without worrying about transferring the receiver.
- A **TTY** (teletypewriter)/**TDD** (telecommunications device for the deaf) is a telephone relay system that is available if you cannot use speech to communicate on the phone; it is used by people who can type. The devices transfer your typed conversation to the person on the other end of the phone who has a TTY or to an operator who reads the message to your listener.
- The telephone company often provides customers with assistive communication devices (speaker phones, phones with large numbers on the buttons, weak-voice amplifiers, and relay systems) free of charge through its deaf and disabled services. A doctor's signature certifying that you have a speech or motor disability is usually required, but if you are unable to obtain such certification, you can purchase these aids through the telephone company or at retail telephone and electronics stores. Consult with your speech pathologist or local ALS Association chapter to see what is offered in your area.


Functional Movement Abilites
Upper Extremity and Head Movement
No Upper Extremity Movement
Locked-In Syndrome
Yes/No System
Letterboard/Picture Board
MegaBee
Writing/White Boards
Personal Computers with Voice Output
Smart Phone Produces (IPad, IPod Touch)
Speech generating Device
Yes/No System
Letterboard/P
Picture Board
MegaBee
Headmouse
Scanning Device
Eye Gaze
Yes/No System
Letterboard/Picture Board
MegaBee
Eyegaze
Hitachi/BCI (Research only)

A B 1 C D	E F 2 G H	I J K 3 L M
N O 4 P Qu	R S 5 T U V	W X 6 Y Z

SPACE - NEW WORD

START OVER

FRONT

EYE GAZE BOARD

1. The partner holds the board at eye level approximately two to three feet from the user
2. The partner should be seated at the user's eye level (while holding the board between him/herself and the user)
3. The user looks at the "dot in the square containing the letter he/she wants
4. The partner will confirm what square the user is looking at by saying that number
5. The partner will then read the letters in that square, pausing after each one
6. When the partner reads the desired letter, the user should indicate yes
7. Follow steps 3-6 until the message is spelled

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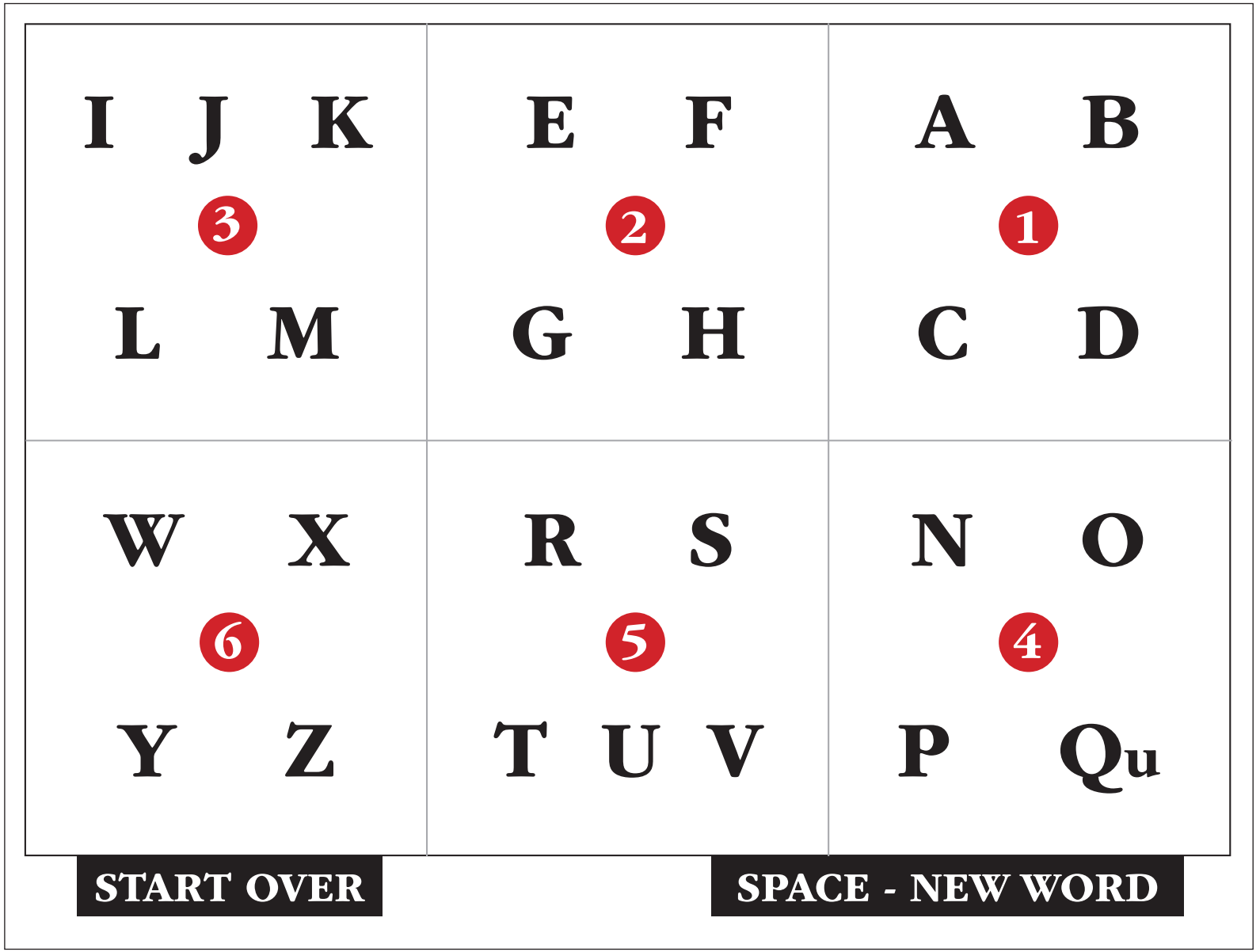

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1. A B C D	5. Q R S T
2. E F G H	6. U V W X
3. I J K L	7. Y Z ? #
4. M N O P	

8. YES 9. NO 10. MAYBE

11. START OVER

12. SPACE - NEW WORD

FRONT

SCANNING

1. Establish a reliable yes response for the user.
2. The partner asks what number row the user's first letter is in. User confirms the numbered row and the partner then reads the letters aloud until the user indicates the correct letter.
3. This pattern continues until the user spell out their message.
4. Upon conclusion of the message, the partner should repeat the entire message to make sure he/she understands.

DIRECT SELECTION

1. Place the board where it is most comfortable for the user to access.
2. The user will point to individual letters in order to spell out a message.
3. The partner must pay close attention to the letters being selected and repeat each letter back to the user for confirmation
4. Upon conclusion of the message, partner should repeat the entire message to make sure he/she understands.

5. Q R S T	1. A B C D
6. U V W X	2. E F G H
7. Y Z ? #	3. I J K L
	4. M N O P

10. MAYBE 9. NO 8. YES

11. START OVER

12. SPACE - NEW WORKD

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How Swallowing Can Be Affected in Patients with ALS

Compiled by: Merisa Palovcak M.S. CCC-SLP/L, Speech-Language Pathologist

Typical Swallowing Problems Associated with ALS

Oral Phase

- Difficulty sucking on a straw
- Inability to remove food from a spoon
- Chewing fatigue/weakness
- Food pocketing or residual food between the teeth and cheeks
- Food "sticking" to the roof of the mouth
- Inability to "gather" food together with the tongue
- Inability to keep mouth closed while chewing
- Drooling
- Food/liquid leaking out of the mouth during chewing and drinking
- Runny nose while eating
- Food/liquid sliding to the back of the mouth before a swallow is initiated

Pharyngeal Phase

- Choking/coughing with foods/liquids
- Need for multiple swallows to clear food through the throat
- Difficulty coordinating breathing and swallowing

Esophageal Phase

There are currently no Stage III problems associated with ALS, however some signs of esophageal dysphagia may include feeling of food sticking in throat/chest, pain made worse with solid foods.

What to look for should you begin to develop difficulties swallowing

1. Notice the time of day the difficulties arise. Is it in the morning, afternoon, or evening?
2. Become aware of the time of the meal the difficulties arise. Is it at the beginning, middle or end of a meal?
3. Note which food consistencies are causing you to have the most trouble. Is it liquids? Is it solid foods?
4. How would you characterize the problem? Are you coughing/choking? Does it feel like you cannot get the food to go down your throat? Is the food sticking in your throat? Is the food difficult to chew?
5. What modifications have you already implemented to make swallowing easier?
6. Are you feeling increasingly fatigued during the course of a meal?
7. Are you becoming Short of Breath?
8. Are you noticing changes in your voice during meals? Does it sound wet or gurgly?

Facilitative and/or Compensatory Strategies Commonly Implemented in Patients with ALS

- Swallowing is optimized if one controls the amount, the texture, and the body/head positioning during feeding. Contact your SLP if you are not able to position upright while eating.
- Control the size of the bite you take: ½ teaspoon size bites, drain mixed consistencies, small single sips of liquids.
- Never eat/drink while lying down. If you must eat/drink while in bed, lay on your side. While this may improve PO (by mouth) management and patient safety when in a reclined position, risk for aspiration is greater. Inform the SLP of any positioning restrictions/needs.
- Pills are often easier to take when they are placed in applesauce or pudding or taken with juices such as natural nectars (pear, apricot) or thicker like tomato juice.
- Follow your body's messages. If chewing is too fatiguing and the portions you are taking are too large, modify your style of eating. At the beginning of a meal, eat the foods that require chewing first as they require more energy. Save the softer foods for the end of the meal as they do not require as much manipulation. Consider smaller meals throughout the day. Also, eat the foods with greater nutritional value first.
- Alternate between liquids and solids to conserve energy by aiding in the propulsion of foods through the throat (pharynx).
- Chin tuck or neck flexed to chest may assist with clearance of food and may increase airway protection. If this technique is needed/used please inform your SLP.
- Use the "Can/Can't" list to help you decide which textures are optimal for you.

Signs and Symptoms of Aspiration (food going down the trachea instead of the throat)

- Coughing/choking when eating
- "Wet" vocal quality
- Eyes and/nose watering or tearing
- Increased respiratory rate with labored breathing
- Persistent low grade fever
- Redding of the face
- Facial Grimace
- Sweating

Pain and ALS

Cramps

- One of the early symptoms and can occur in over 80% of the ALS population. PALS have reported cramps in the muscles of their limbs, abdomen, back, neck, jaw and even tongue.
- Described as a sudden, involuntary sustained muscle contraction that can interrupt activity and sleep.
- Generally limited to a certain muscle group, e.g., hands or feet.
- More likely to occur with over-exercising a muscle group, if the muscle is cold, or if circulation is decreased.
- Treatment for cramps includes stretching and massaging the affected area, drinking tonic water, and the prescribed medications Neurontin®, Dilantin® and Tegretol®.

Spasticity

- Feels like a sustained pulling or tightening of a muscle, and can be quite severe.
- All of the voluntary muscles can be affected by spasms but tends to affect larger areas of the body, e.g., arms, legs, trunk.
- Heat and cold may help to decrease the spasm, along with range of motion and stretching exercises.
- A physical or occupational therapist can teach the PALS and/or caregiver how to do the exercises and treatments.
- Baclofen®, Zanaflex®, Clonazepam® and Botox injections may also be beneficial

Numbness or a burning sensation of the legs, feet and hands.

- Elevating the limb may help
- Prescribed medication, Neurontin®.

Immobility can cause pain along pressure or bony areas, e.g., the sacral area, heels, elbows and hips.

- Change position ideally every two hours or have someone change position for them.
- Use of gel pads and mattresses, sheepskins and/or “egg-crates” are often helpful in relieving pressure. Gel pads and mattresses are usually covered items under most insurance plans. Sheepskins and egg-crates can be purchased at Medical Supply stores.

Joint contractures

- Occur when joints have little or no mobility, secondary to muscle death surrounding the joint. Simply stated, a joint contracture is a permanently immobile joint.
- Any voluntary or involuntary movement of the joint can cause severe pain.
- Particular concern with the ALS population are the shoulder joints. Once these joints are contracted, it is often difficult to transfer a PALS pain-free.
- Contractures are preventable with daily range of motion exercises and/or splinting the affected joint(s).

Fatigue and ALS

A common symptom of ALS is fatigue. Fatigue occurs in ALS patients because of decreased muscle strength—muscles affected by ALS become fatigued from normal use. In addition, the muscles used to compensate for weakened muscles can become fatigued from overuse. The ALS patient who has a weakened left leg will put more weight on the right leg, resulting in fatigue in both limbs.

Fatigue is more than feeling tired—it is often described as extreme exhaustion by ALS patients. **Energy conservation is the treatment for fatigue**. Principles of energy conservation include:

- Balancing rest and activity. Save your energy for things you enjoy doing.
- If someone is available to assist you with mundane tasks like cleaning, accept the help.
- Do not try to push through the fatigue – it will not make it easier the next time.
- Use Assistive Devices such as tub chairs, back scrubbers, thick-handled eating utensils, etc. An Occupational Therapist can determine what assistive devices would be best for you.
- Get a handicapped parking sticker. Your local department of motor vehicles and/or the ALS Chapter Social Worker has the form for this.
- Try to establish a regular sleeping pattern.
- Avoid stressful situations as much as possible. Stress is fatiguing. Find pleasant, relaxing activities that work for you and do them.
- Organize/prioritize/plan ahead.
- Use common sense. If you have trouble walking, don't resist getting a motorized wheelchair. If you have difficulty speaking and are having company in the evening, save your voice for when you want to be able to speak as well as possible.

Although in ALS you can't take away fatigue, by planning ahead you can still participate in activities you enjoy.

Exercise and ALS

The physical or occupational therapist can make recommendations for exercise based upon each patient's specific needs and abilities.

- **Strengthening exercises** are **not generally recommended** for patients with ALS as research has shown them to be ineffective in maintaining or increasing strength once muscles have been affected by the disease.
- **Stretching and Range of motion (ROM) exercises are done** to maximize flexibility and mobility of the joints on which they are performed. Joints maintain their normal range of motion by being moved. It is therefore very important to move all joints every day. Stiff joints and shortened muscles can cause pain and can make it hard to do normal daily activities.
- The therapist can **instruct you or your caregiver in a home ROM program** that you can follow through with daily. It is important to remember that as the disease progresses, the type of ROM exercises needed may change.
- It is important to realize **that these exercises will not strengthen muscles that have been weakened by ALS.** Once the supply of motor neurons that control a particular muscle has degenerated, it cannot be regenerated by exercise. Trying to excessively exercise muscles that are weak already will only increase weakness and rob energy that is needed for daily routines and activities that are enjoyed.
- **Monitor Fatigue Level**
- **Stop** if causing pain, cramps, or fasiculations
- **Avoid Eccentric contractions**
- **Avoid resistance and high repetitions**

Passive Range of Motion Exercises

General instructions:

- Ideally, these exercises should be done at least once per day.
- Doing the exercises after a shower or 20 minutes of heat, can improve the elasticity of the muscles and increase the effectiveness of the stretching.
- Do each exercise 3-5 times. Move to the point of resistance and hold for 20-30 seconds.
- Keep limbs supported throughout motion.
- Move slowly, watching the patient's face for response to ROM.
- The eventual goal is full range of motion. This does not have to be achieved on the first repetition.

ALS and Cognitive Changes: A Guide for Patients and Families

Overview

A link between ALS and cognitive dysfunction (problems with thinking and/or behavior) was first noted in the late 1800's. However, it's only been in the last few years that new research has shed important light on how these two conditions might be related. What was once considered a rare link between two different diseases has now been identified as a more common combination. Knowledge about the relationship between ALS and cognitive changes is growing and changing. More research is planned to increase medical understanding and to help in the management of patient care.

There is a very wide range of what patients with ALS may experience — from no cognitive changes at all, to mild or moderate difficulties, and in some cases, more severe problems.

Many people with ALS will have no changes in behavior or thinking.

Recent studies suggest that it is not uncommon for patients with ALS to have mild-to-moderate thinking or behavioral problems. In some cases, the thought or behavior problems may be so mild and subtle that they aren't noticed by friends or family members and don't interrupt daily activities, and are only recognized through neurologic testing. For others, the symptoms (described below) are more noticeable and do impact daily activities.

A sizable minority of people with ALS may have a more serious form of these problems. It's called frontotemporal lobar dementia (FTLD). The location of the degeneration of neurons that causes the problems happens in the front and side parts of the brain.

Because for a long time it was believed that ALS did **not** affect a person's mental processes, health care professionals are now taking another look at some commonly-held approaches to managing the care of people with ALS. For patients and families, just knowing that problems with cognitive function and behavior may be a part of the ALS disease process can be helpful to explain what they may already be observing. There are a wide range of approaches families can take to better manage problems related to these problems. It's important to know that the cognitive changes that can be experienced with ALS are do to degeneration of brain cells, and are not a psychiatric illness.

What Are Cognitive Impairment (CI) and FTLD?

Cognitive Impairment (CI) and Frontotemporal Lobar Dementia (or FTLD) are a mild to severe continuum of a brain disease that causes particular brain cells to slowly, consistently die. This illness causes the patient to have personality changes, language difficulty and/or behavioral disturbances. Some people with ALS show only limited signs of cognitive or behavioral changes (Cl), while others develop symptoms that may lead to a diagnosis of FTLD. The neuron degeneration and changes of CI and FTLD have been identified using Magnetic Resonance Imaging (MRI) technology.

The behavior and thinking problems experienced in the more severe FTLD are different from the dementia of Alzheimer's disease and presents different signs and symptoms. For example, people with FTLD typically develop *behavior* symptoms first, while people with Alzheimer's disease usually develop *memory* problems first. Many of those with FTLD may not experience memory problems at all.

Symptoms of CI and FTLD

Sometimes the symptoms of ALS can be confused with symptoms of CI or FTLD. Medications, depression, or problems with respiratory function can all cause some of the same symptoms that CI or FTLD can cause. It's important for a patient's symptoms to be evaluated by an ALS health care provider to find out the cause of the problems. Because people with ALS typically experience a steady decrease in their ability to speak, swallow, move and perform activities of daily living, it can be easy to miss noticing some common thinking problems.

The main symptoms of CI and FTLD have to do with behavior and personality changes and they develop gradually over time. Some changes in personality might include the following:

- Less sensitive to the needs of others
- Behave in ways that are embarrassing, inappropriate, and uncharacteristic
- "Too cheerful" and may seem childish
- More withdrawn and need prompting to do most things
- Changes in behavior such as eating lots of sweets and stuffing food in their mouth without swallowing
- Fixated on one idea or activity
- More aggressive

Other symptoms of CI or FTLD have more to do with thinking. For example, many people with cognitive impairment may not recognize that they have any problems or changes, and as a result make some bad decisions about how to take care of themselves, perform their work, or manage their health. They may have difficulty solving problems, especially if the problem is something new such as using a new piece of equipment, or if it involves more than a few steps.
Some people lose insight into their illness, and so would not be able to discuss the cognitive and emotional changes they are experiencing.

Patients may not care much about or recognize their problems, they may not be able to focus attention, or they may become overly agitated. 'Executive function' is a thinking skill that is difficult for ALS patients with cognitive problems. 'Executive function' refers to a type of complex thinking when people handle many types of information at the same time; such when one drives a car through busy streets at rush hour. This skill is also used when patients are asked to make complex decisions about life-style changes and medical interventions when the patients is given detailed information about changes in speaking, swallowing, breathing, and muscle control.

Each patient with these cognitive changes will have different kinds of problems.
Some with only changes in behavior and personality,
Some with only thinking problems, and
Others with both types of problems.

Because of their thinking problems, some patients are not able to understand the changes that are happening to them. Family members too may see the cognitive changes as just frustration or anger with the illness itself or with the caregiver.

Risk Factors for CI and FTLD

Patients over the age of 60, who had bulbar onset ALS, poor breathing, or a family history of dementia, may be more likely to have cognitive impairment or FTLD.

Diagnosis of CI and FTLD

A diagnosis of CI or FTLD is made when particular behaviors are seen in patients. People with FTD may have problems interacting with others socially, have less emotional reactions to things that would normally upset people, and they often do not understand what is happening to them.

To diagnose the presence of this brain disease, your physician will refer you to a neuropsychologist for a neuropsychological and neurobehavioral evaluation. This evaluation will take several hours and are not blood tests or like other physical diagnostic tests. In the ALS clinic, a few tests may be given to patients to see if they should have this type of evaluation. Patients may be asked to say a list of words as quickly as they can and caregivers may be asked a series of questions about the patient's behavior.

Learning that the Patient Has Cognitive Changes

If doctors suggest that Cl or FTLD is present, the diagnosis is sometimes a relief to the family, because it often explains problems with thinking or behavior that were previously misunderstood. Some family members say that learning more about CI and FTLD helps explain behaviors such as aggressive behavior or a patient's lack of concern about what is happening to them or their families. Naturally, this can be a very difficult time for patients and family members.

Things that can help patients and families cope with the thinking and behavior changes the patient may experience, and may be able to make the situation a bit easier:

Become informed about what is happening.

Talk frankly with the patient's health care providers about the symptoms, possible treatments and ways to help the patient. Ask about things caregivers and family members can do to help the specific thinking or behavior problems their loved one is having.

Get support or counseling.

Know that developing cognitive changes have nothing to do with anything the patient or family may have done, or not done.

Length of Survival and Acceptance of Medical Treatments

Studies suggest that, for reasons that are not yet completely understood, patients with both ALS and CI or FTLD may have a shorter lifespan than those patients with ALS alone. Some health care providers believe that this problem is related to their observations that ALS patients with cognitive and behavioral problems do not follow their doctor's recommendations as often as patients without CI or FTLD.

This shortened life span may affect families, especially as they involve patients in treatment decisions and in legal matters, such as advance directives, wills, and related documents. These challenges may occur sooner than family members had anticipated and the loved one with ALS may not want to participate in or accept the important decisions that need to be made.

Recommendations for Care and Support

Ways you may be able to help those with cognitive or behavioral problems:

Remember that the patient has these problems because their brain is changing. The patient can not simply 'fix' the problems by trying harder. Equally important, it is not the caregiver's fault. The caregiver and family can't 'fix' the problem by trying to work faster or harder. Caregivers and families are not to blame if patients' behavior or thought problems worsen or interfere with socialization and daily activities.

Use simple and straightforward language and communicate clearly and directly.

Supervise the patient's eating more closely. Those with poor swallowing may have trouble following medical advice to limit solid foods, or they may place too much food in their mouth at one time. Some patients may eat more food than they need.

Help your loved one make decisions about their care. The patient may not understand why they need to use walkers, wheelchairs, breathing equipment or feeding tubes.

Supervise walking. Patients may make decisions too quickly, without remembering to be careful. They may walk in dangerous places, walk too far, or forget to use equipment such as a walker.

Try to build an atmosphere of comfort and love, with a calm, structured and orderly environment.

Medications for FTLD with ALS:

While physicians cannot cure these cognitive and behavioral problems today, some medicines may provide a degree of relief from symptoms. Because each patient has their own, unique type of problems, your doctor will choose which medicine/s will be helpful for each individual patient.

Summary

For years, patients and families have taken some level of comfort from the thought that although ALS poses unique and serious physical challenges, the one issue they were not likely to have to face was a decrease in cognitive function. Unfortunately, research now suggests that this may not be true for all ALS patients.

The diagnosis and treatment for Cl and FTLD with ALS is improving. Researchers are hopeful that the discovery of the connection between ALS and Cl and FTLD among some patients may speed the understanding of the cause of ALS and provide important clues to the puzzle ALS still presents.

In applying any of this information to the care of a particular individual, we recommend talking with their healthcare professional first,

Lots a Helping Hands and the Care Connection Program

Do you have neighbors, family members, friends ask, "What can I do to help?" Both the National ALS Association and the Greater Philadelphia Chapter suggest that you **put these folks to work**. One way to organize a group of helpers is to utilize a free, secure web community called "Lots a Helping Hands." This website was created in 2005 by caregivers who recognized the need for a way to coordinate care for any family struggling with an ill loved one. Since then, a specific website for persons with ALS (PALS) has been created, as follows: www.alsa.lotsahelpinghands.com

Anyone can create a web community of caregivers, although it is recommended that the person who creates and "coordinates" the care be someone who is close to the PALS, but not the primary caregiver (he or she has enough to do!). The Coordinator would need to meet with the PALS and primary caregiver (CALS) to determine:

- List of tasks that would be of help to the PALS/CALS, such as running errands, providing meals, rides, shopping, laundry, pet care, help with childcare, housework, yard work, providing companionship, and possibly physical help with the PALS.
- List of family members, friends, neighbors, community/church members, called "Participants, who the PALS/CALS would like to be included in their "circle of care" or community of caregivers.

Next, the Coordinator would set-up a (private) web community by creating a team name, e.g., "Nancy's Buddies, listing folks who have been identified by the PALS/CALS, along with a schedule of requested tasks or assignments. It is then the responsibility of the Participants to check the website from time to time, and sign up for tasks. The website automatically notifies the Participant of their assignment one month, one week and one day prior to the scheduled day.

The Care Connection Program is the National ALS Association's name for the same idea/goal of the website—organizing help for a PALS and their loved ones. It is based on the book, Share the Care—How to Organize a Group to Care for Someone Who is Seriously Ill, written by Cappy Capossela and Sheila Warnock (1995). With the Care Connection program, however, a Coordinator may or may not utilize the website depending on their computer comfort level. For example, some Coordinators prefer using the phone to contact Participants. The Care Connection Program also emphasizes a need to limit outside communication to the PALS/CALS so that Participants would only contact the Coordinator, and not the PALS/CALS directly.